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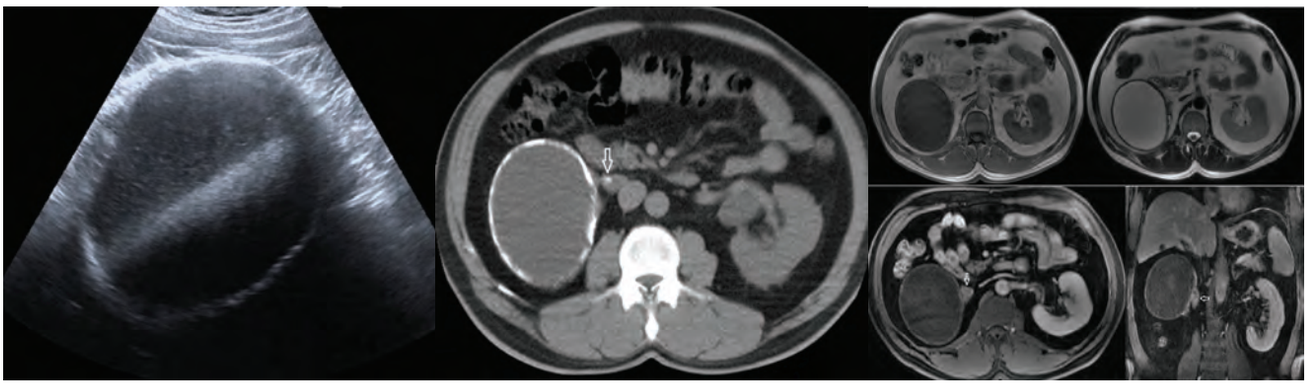
A Rare Cause Of Renal Atrophy: Subcapsular Collection Presente As A Huge Perirenal Complex Cyst

Kılınçer A, Ünal E, Yıldız O



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Orhan Yücel
GATA Göğüs Cerrahisi. AD. 06118, Etlik, Ankara Turkey.
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National Editorial Board

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B. LETTERS TO EDITOR

Hip Ultrasonography in Classification of Proximal Focal Femoral Deficiency: Two Case Reports

Proksimal Fokal Femur Yetmezliği Sınıflamasında Kalça Ultrasonografisi: İki Olgu Sunumu

Proximal Focal Femoral Deficiency

Ümit Yaşar Ayaz[1], Sevin Ayaz[2], Mehmet Ercüment Döğen[1], Baki Hekimoglu[3]
[1]Department of Radiology, Mersin Women's and Children's Hospital, Mersin,
[2]Department of Medical Imaging Techniques, Toros University, Vocational School; Department of Nuclear Medicine, Mersin State Hospital, Mersin,
[3]Department of Radiology, Dışkapı Yıldırım Beyazıt Training and Research Hospital, Ankara, Turkey

*Presented in 22nd International Congress of Radiology, July 1-5, 2002 in Cancun, Mexico.

Özet

Proksimal fokal femur yetmezliği (PFFY) bulunan iki infantta femur başlarını sonografik olarak göstermeyi ve ultrasonografi (US) katkısı ile PFFY sınıflaması yapmayı amaçladık. Sağ uyluğunda kısalık bulunan yedi aylık erkek olgunun (Olgu 1) radyografisinde sağ femurun proksimal yarısında belirgin hipoplazi saptandı. Aynı taraf asetabulum displazikti. US'de femur başı hipoplazikti. Sağ uyluğunda kısalık bulunan 16 günlük erkek olgunun (Olgu 2) radyografisinde sağ femurun sola kıyasla daha kısa ve ince olduğu görüldü. Aynı taraf asetabulumun gelişimi yeterliydi. US'de femur başı normaldi. US, PFFY'li infantlarda kıkırdak femur başlarının durumunu göstererek Olgu 1 ve Olgu 2'nin, sırasıyla Aitken sınıf C ve A olarak sınıflandırılmasına yardımcı olmuştur. US, radyografi ile birlikte PFFY sınıflamasında ve diğer kalça eklemini değerlendirmede kullanılabilir.

Anahtar Kelimeler

Süt Çocuğu; Ultrasonografi; Kalça Eklemi; Femur; Alt Ekstremite Deformiteleri, Doğumsal

Abstract

Our purpose was to sonographically demonstrate the femoral heads of two infants with proximal focal femoral deficiency (PFFD) and to classify PFFD with the contribution of ultrasonography (US). On the radiograph of Case 1, a seven-month-old male with a short right thigh, there was marked hypoplasia of the proximal half of the right femur. Ipsilateral acetabulum was dysplastic. On the US, the femoral head was hypoplastic. On the radiograph of Case 2, a 16-day-old male with a short right thigh, the right femur was shorter and thinner than the left femur. An adequate ipsilateral acetabulum was found. US revealed a normal femoral head. US helped the classification of Case 1 as Aitken class C and Case 2 as and Aitken class A, by demonstrating the status of cartilaginous femoral heads in PFFD. Combined with radiography, US can be used in the classification of PFFD and in the evaluation of the unaffected hip.

Keywords

Infant; Ultrasonography; Hip; Femur; Lower Extremity Deformities, Congenital

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Corresponding Author: Ümit Yaşar Ayaz, Mersin Women's and Children's Hospital, Department of Radiology, Mersin, Turkey.
T.: +90 3242230701 GSM: +905377639442 F.: +90 3242230722 E-Mail: umityasarayaz@gmail.com

Introduction

Proximal focal femoral deficiency (PFFD) is a rare developmental disorder of the proximal segment of the femur and the acetabulum, resulting in a shortening of the affected limb and ensuing functional impairment. The incidence of PFFD was reported as one per 52,029 of the population (0.002%) [1]. According to the radiological classification proposed by Aitken [2], femoral head is present in class A PFFD, with an adequate acetabulum and a very short femoral portion. Initially, there is no bony fusion between that portion and the femoral head in Aitken class A PFFD. However, through skeletal growth and maturation, a bony fusion develops between the diaphysis of the femur and the head, neck, and trochanteric component. In Aitken Class C PFFD, the acetabulum is seriously dysplastic; the diaphysis of the femur is short with a tapered upper part; the femoral head is absent or very small with no connection with femoral diaphysis; and the femoral head (if present) never gets ossified [2]. Our aim in presenting the following two case reports was to ultrasonographically demonstrate the femoral heads of infants with PFFD, to present their plain radiography findings, and to classify them with the contribution of US data. Aitken's classification [2] was used as the reference.

Case Report 1

The male infant was seven months old on admittance, born at term after an uneventful pregnancy. No maternal risk factors including gestational diabetes or exposure to teratogenic agents during pregnancy were reported by the parents. Family history was unremarkable and there was no consanguinity. The physical examinations of the parents, including their limbs, were normal. The patient's parents were informed about the examination procedures and consent was obtained from them. All procedures were performed according to the World Medical Association Declaration of Helsinki (revised in 2000, Edinburgh). On physical examination of the infant, his right thigh was shorter than the left one and was bulky. The left leg, upper limbs, and face were normal. Physical examination, routine laboratory tests, and bone density evaluations revealed no systemic disease or rickets. On plain radiograph there was marked hypoplasia of the proximal 1/2 portion of the right femur with a tapered proximal end. The distal 1/2 portion of the right femur was thinner and shorter than that of the contralateral femur. The right acetabulum was dysplastic. There was a small notch superior to the right acetabulum, thought to represent a pseudoacetabulum formation. There was some superior displacement of the right femur; its tapered proximal end pointed to the pseudoacetabulum formation. On the left side, the acetabular angle was 41°, representing a relatively shallow acetabulum. Tibias and fibulas were present on both sides and no skeletal abnormality was observed in the left leg (Figure 1). On US images obtained by a 7.5 MHz linear-array transducer, a hypoplastic cartilaginous femoral head was demonstrated on the right side. Its diameter was 13.4 mm with no ossification center, while the left femoral head with an echogenic ossification center had a diameter of 19 mm. During the Barlow maneuver, US demonstrated that the right femoral head was superiorly displaced and located in an acetabulum-like depression on the iliac bone (Figure 2), concordant with the pseudoacetabulum

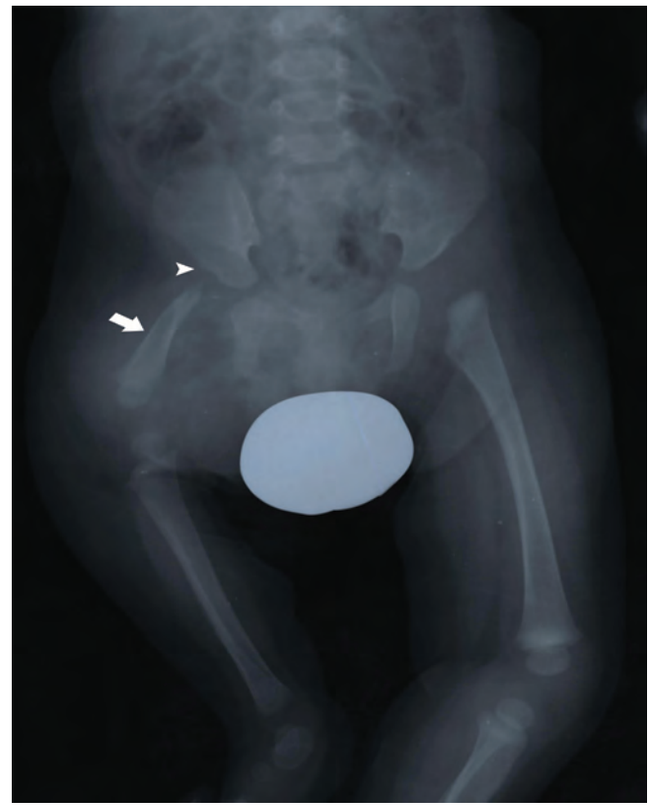

Figure 1. Anteroposterior plain radiograph of Case 1. Short and thin right femur with a tapered proximal end (white arrow). Ipsilateral acetabulum is dysplastic. Small notch superior to the right acetabulum was thought to represent a pseudoacetabulum (white arrowhead). Tibias and fibulas are present on both sides.

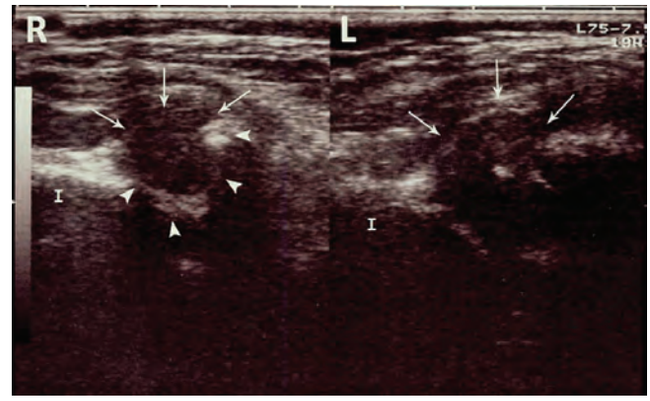

Figure 2. Coronal US views of both hips in Case 1 during Barlow maneuver (R: Right side, L: Left side, I: Iliac bone). Smaller cartilaginous femoral head on the right side (white arrows) with no ossification center and normal-sized femoral head on the left side (white arrows) with a tiny echogenic ossification center. Right femoral head is located in a notch on iliac bone, concordant with a pseudoacetabulum (white arrowheads), in which no hypoechoic Y cartilage could be demonstrated.

detected on plain radiograph. The left femoral head coverage (FHC) was 50% in neutral position and decreased to 38% during the Barlow maneuver. The presence of a dysplastic acetabulum with an ipsilateral hypoplastic cartilaginous femoral head that was lacking an ossification center suggested that the PFFD in the present case had features consistent with Aitken class C.

Case Report 2

The male infant was 16 days old on initial admittance. He was born at term after an uneventful pregnancy. His nonconsanguineous parents reported no maternal risk factors including gestational diabetes or exposure to teratogenic agents during pregnancy. Family history was unremarkable. The physical ex-

aminations of the parents including their limbs were normal. The patient's parents were informed about the examination procedures and gave their consent. All the procedures were performed according to the World Medical Association Declaration of Helsinki (revised in 2000, Edinburgh). On physical examination of the infant, his right thigh was shorter than the left one and was bulky. The left lower limb, upper limbs, and face were normal. His physical examination and routine laboratory tests revealed no systemic disease or rickets. On his first plain radiograph obtained on initial admittance, his right femur was shorter and thinner than the left femur. An adequate right acetabulum was found to be similar to the left acetabulum. There was a separate ossification center in the proximal portion of his right thigh representing the femoral neck (Figure 3), which was seen to have enlarged and to have fused with the more distal portion of the right femur four months later (Figure 4). Tibias and fibulas were present on both sides and no skeletal abnormality was observed on the left leg. On the last control radiograph obtained 12 months after the initial one, ossification centers of both femoral heads were visible and rather symmetrical. Right femoral length and thickness gradually increased and substantial improvement was observed. The right acetabular angle was 16° and the left acetabular angle was 30°, both within normal ranges (Figure 5). The first US examination performed by a 7.5 MHz linear-array transducer on initial admittance revealed a normal cartilaginous femoral head with no ossification center on the right side. Its diameter was 15.4 mm, close to the diameter of the left femoral head which was measured as 15.9 mm.

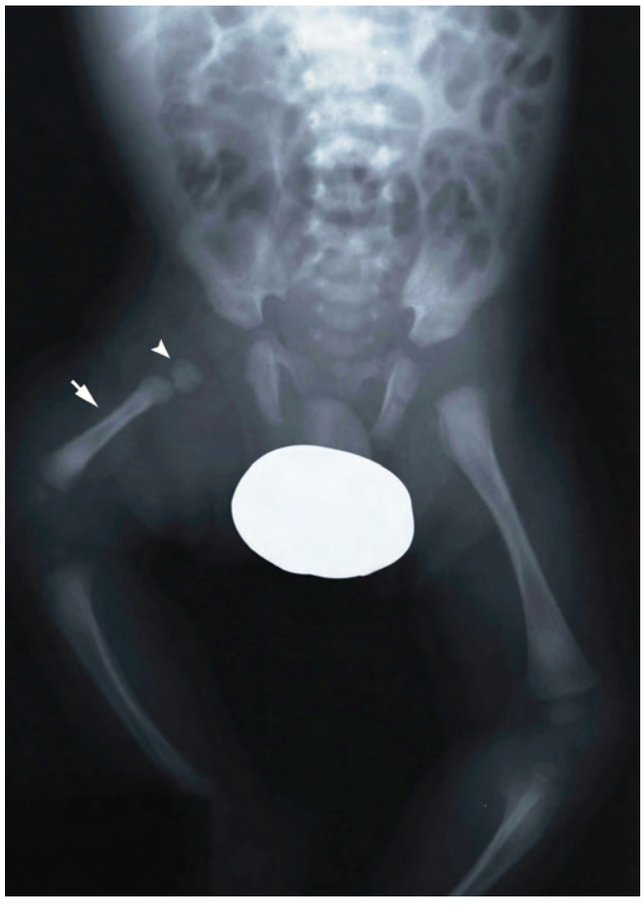

Figure 3. Anteroposterior plain radiograph of Case 2 on initial admittance. Right femur is short and thin (white arrow). Tibias and fibulas are present on both sides. A separate ossification center in more proximal portion of his right thigh representing the femoral neck is visible (white arrowhead).

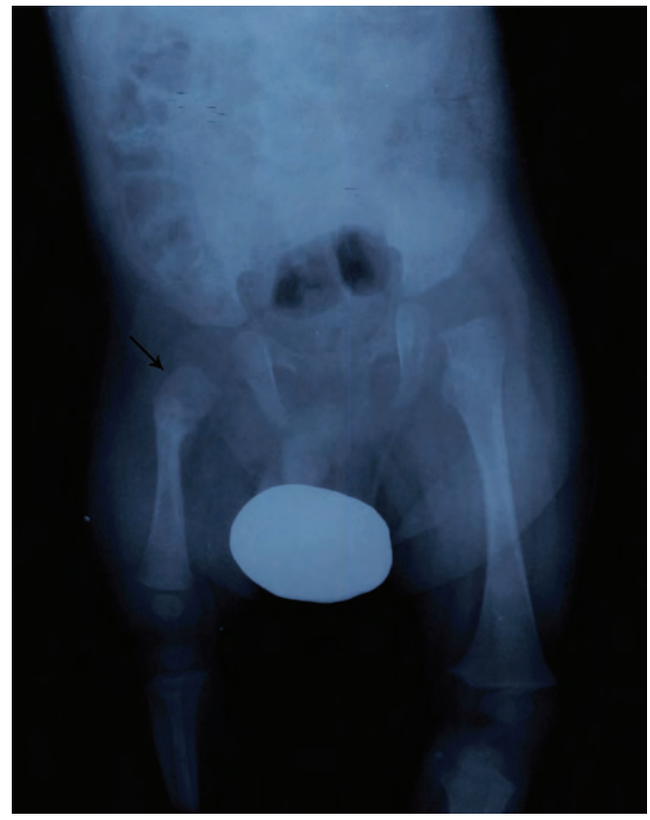

Figure 4. Anteroposterior radiograph of Case 2, four months after initial radiograph. Enlarged ossification center (black arrow) is fused with more distal portion of the right femur. An increase in length and thickness of right femur is observed.

During the Barlow maneuver, the right FHC was 22.6% (Figure 6), concordant with marked subluxation and the left FHC was 31%, higher than the right one but also concordant with subluxation. With the Ortolani maneuver both femoral heads returned

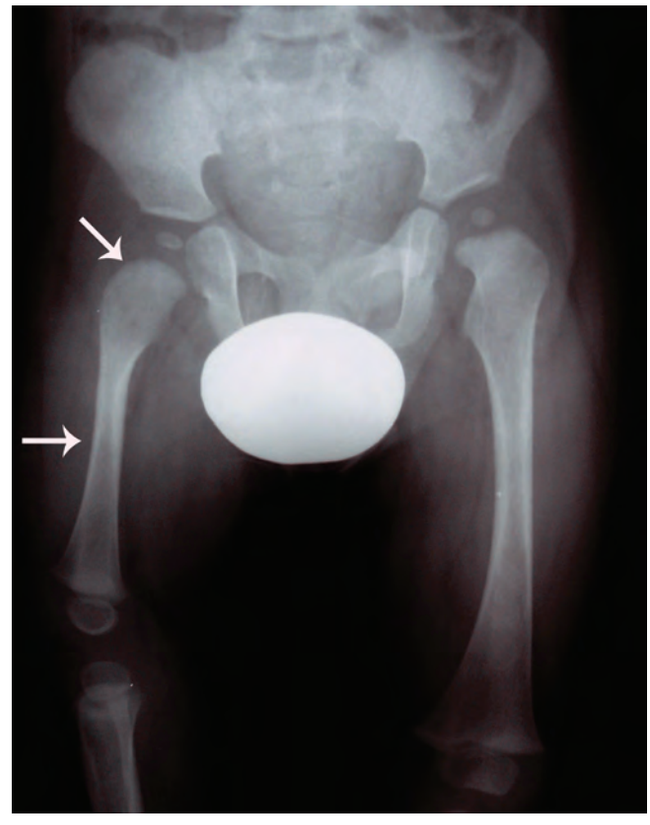

Figure 5. Anteroposterior radiograph of Case 2, 12 months after initial radiograph. Ossification centers of femoral heads are visible on both sides. Substantial improvement in development of right femur is observed (white arrows). Acetabular angles are within normal range on both sides.

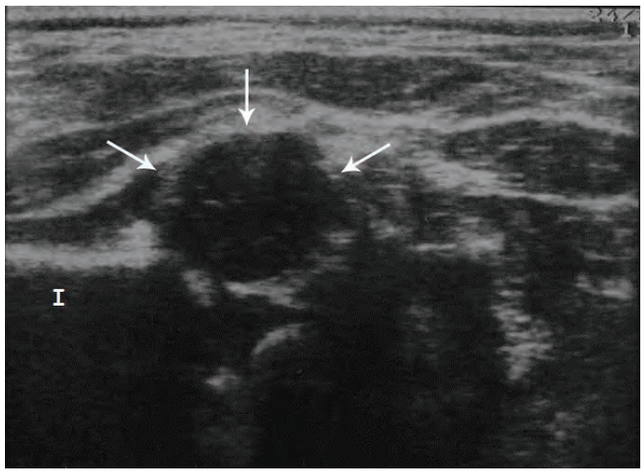

Figure 6. Coronal US view of the affected hip (right) in Case 2 during Barlow maneuver, obtained on initial admittance. Decreased femoral head coverage (FHC) and marked subluxation of cartilaginous femoral head (white arrows). I: Iliac bone.

to their resting positions. With the contribution of initial US findings, Case 2 was classified as Aitken class A. Four months later, control US revealed marked increase in FHC on both sides without any sign of overt subluxation, the right one being 48.2% (Figure 7) and the left one being 47.5% during the Barlow maneuver. On the last control US performed 12 months after the initial one, the right FHC was 49.2% (Figure 8) and the left FHC was 51% during the Barlow maneuver. Overt subluxation could not be detected on both sides. Eventually, follow-up data helped confirm the initial classification of the case as Aitken class A.

Discussion

The term PFFD is applied to a spectrum of conditions characterized by partial absence and shortening of the proximal femur(s). The pathology is mostly sporadic but familial PFFD cases have also been reported [3]. Aitken's classification [2] is most widely employed in both diagnosis and therapeutic planning. Radiological features of PFFD are present at birth. Except in the most severe cases, the distal end of the femur is usually normal. In milder forms (classes A and B), the femoral head and neck ossify and fuse, but a non-progressive subtrochanteric varus deformity is constant. In more severe types, the acetabulum and the femoral head are absent, an iliac bony projection is observed lateral to and above the dysplastic acetabulum, and the femoral diaphysis is either rounded, pointed proximally, or almost completely absent. In all four forms, the femur is prominently shortened. After age two, evaluation of plain radiographs and appropriate assignment into one of the four Aitken's classes become more feasible and more definitive than in infants below this age. The growth ratio of the abnormal to the normal limb throughout childhood is usually constant [4]. The Amstutz classification [5], another common classification system, has also been used in PFFD and has been applied to MRI [6].

To compensate for the inability of radiographs to demonstrate cartilage, MRI and US have been used to obtain more data in cases with PFFD [6–8]. Kayser et al. [7] reported that with US, the iliac line, femoral head, and greater trochanter could reliably be visualized in their patients with PFFD. In our cases, both the acetabuli and cartilaginous femoral heads could be evaluated using US, which helped us in classification of PFFD. Case 1 was a delayed case and the formation of a small pseudoacetabulum on the right side was thought to be due to a long-standing pressure effect of superiorly dislocated hypoplastic right femoral head on iliac bone. Though Case 1 had features consistent with Aitken class C, clinical follow-ups, control radiographs, and US examinations were recommended to the parents to verify the initial classification. Contact with the patient could not be established after the first examination, so we could not re-evaluate him. In Case 2, control US examinations revealed a normal-sized cartilaginous femoral head in its normal position on the affected side. We concluded that these features of Case 2 represented Aitken class A deformity, the mildest form of PFFD. Maldjian et al. [6] used MRI in diagnosis and classification of PFFD, stating that MRI was more accurate than radiographic evaluation for the classification of PFFD, particularly prior to the ossification of cartilaginous components in the femurs. MRI has perfect contrast resolution and it is superior to US in demonstrating all the bony elements (femoral

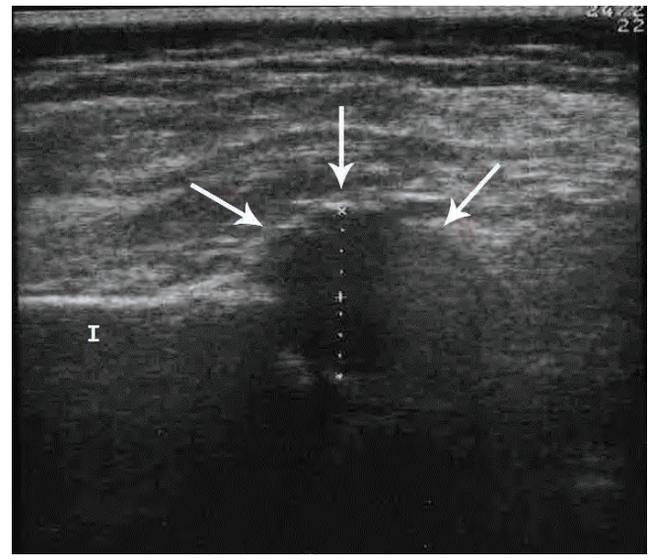

Figure 7. Coronal US view of the affected hip (right) in Case 2 during Barlow maneuver, obtained four months after the first US examination. Marked increase in FHC (calipers) without any sign of overt subluxation (white arrows). I: Iliac bone.

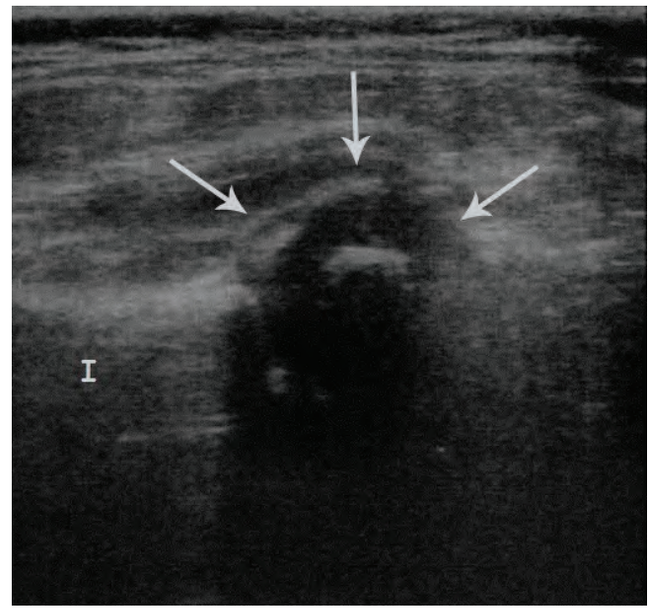

Figure 8. Coronal US view of the affected hip (right) in Case 2 during Barlow maneuver, obtained 12 months after the first US examination. Ossification center with posterior acoustic shadowing is visible in the center of femoral head. Marked increase in FHC without any sign of overt subluxation (white arrows). I: Iliac bone.

head, femoral neck and shaft, acetabulum, iliac bone) and cartilage together, in clearly depicting their relationship with each other, and in showing any impingement [8], regardless of any limiting factor experienced with US, such as adverse effects of tissue thickness and acoustic shadowing caused by bone. However MRI is more costly than US and sedation is required in infants. Among all the imaging modalities, US seems to be the most practical method to evaluate non-ossified femoral head in PFFD. Like MRI, US is also free of ionizing radiation, but unlike MRI, it does not require sedation and it is relatively less costly. US also provides real-time images during maneuvers applied in dynamic hip examination. US is particularly useful as a rapid and practical tool in the very first diagnosis and classification of PFFD, prior to detailed MRI examinations which are stressful for the infant and the family. But experience in assessment of infant hips with US is essential; otherwise, misdiagnosis would be inevitable due to drawbacks in imaging. In our cases, combined with the radiographs, US was sufficient to evaluate the existence, size, and position of cartilage femoral head, both on the affected and unaffected sides. But we consider that MRI and US can be performed together, particularly in indeterminate cases, to combine the advantages of both techniques and to more confidently make the classification.

In conclusion, US was useful both in demonstration of cartilaginous femoral heads and in evaluation of their diameters/positions in our two infants with PFFD. Combined with plain radiography findings, US can be used in the classification of the affected femur and in evaluation of the unaffected side to rule out the coexistence of a possible developmental hip dysplasia.

Competing interests

The authors declare that they have no competing interests.

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Case Report

JCAM

A Rare Cause of Snoring: Isolated Nasopharyngeal Lymphangioma

Horlamanın Nadir Bir Nedeni; İzole Nazofarengeal Lenfajiom

Izole Nazofarengeal Lenfanjiom

Hulya Eyigor[1], Oguzhan İlden[1], Dinc Suren[2], Dondu Nergis[2], Levent Renda[1]
[1]Department of ENT Head and Neck Surgery, [2]Department of Pathology,
Antalya Education and Research Hospital, Antalya, Turkey

38. Türk Ulusal Kulak Burun Boğaz ve Baş Boyun Cerrahisi Kongresi 2016'da Poster olarak sunulmuştur.

Özet

Lenfanjiomalar, lenfatik sistemin nadir konjenital tümörleridir. En sık çocukluk çağında görülmekte ve sıklıkla yerleşim yeri baş ve boyun bölgesidir. Nazofarenksin izole lenfanjioma tululumu oldukça nadir olup literatürde bildirilmiş bir kaç vaka bulunmaktadır. Bizde bu çalışmamızda horlama ve sağ kulakta tıkanıklık yakınmasıyla polikliniğimize başvuran ve izole nazofarengeal lenfanjioma tanısı alan 40 yaşında bayan hastayı literatür bilgileriyle sunduk.

Anahtar Kelimeler

Lenfanjioma; Nazofarenks; Horlama; CD31; CD34

Abstract

Lymphangiomas are rare congenital tumors of the lymphatic system. They are most often seen in childhood and frequently are located in the head and neck region. Nasopharynx involvement of an isolated lymphangioma is extremely rare, with very few cases reported in the literature. Together with the relevant information in the literature, we present here a case of a 40-year-old female who presented at our polyclinic with complaints of snoring and obstruction in the right ear and was diagnosed with isolated nasopharyngeal lymphangioma.

Keywords

Lymphangioma; Nasopharynx; Snoring; CD31; CD34

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Corresponding Author: Hulya EYIGOR, Department of ENT, Antalya Research and Education Hospital, 07100, Muratpaşa, Antalya, Turkey.
GSM: +905372169753 F.: +90 2422494462 E-Mail: hulinar@yahoo.com

Introduction

Lymphangiomas are rarely seen congenital tumors of the lymphatic system. They are most frequently seen in childhood and generally diagnosed in infancy; approximately 90% of cases are seen by the age of 2 years [1]. According to the size of the cavity, lymphangioma can be classified as microcytic, macrocytic, or cystic hygroma. These tumors are seen most often in the head and neck region and more rarely may be observed in the axilla and abdomen. As isolated nasopharyngeal lymphangiomas are extremely rare, few cases have been reported in the literature since the first case in 1966 [1,2].

In the light of information in literature, we present the case of a 40-year-old patient diagnosed with isolated nasopharyngeal lymphangioma, who underwent surgery and had no complaints during a 1-year follow-up period.

Case Report

A 40-year-old female presented at the polyclinic with complaints of intermittent obstruction of the right ear and snoring which had been ongoing for one year. In the endoscopic nasal examination, a polyposis mass was determined originating from the right posterolateral nasopharynx and extending to the oropharynx, narrowing the nasopharyngeal cavity (Figure 1). The neck examination and other ENT examinations were normal. No additional developmental anomaly was determined in the patient.

On the computed tomography (CT) examination, evident thickening was determined in the torus tubarius in the nasopharynx right hemisphere and obliteration in the Rosenmüller fossa. On magnetic resonance imaging (MRI) of the nasopharynx, a mass was determined, 16 x 12 x 25 mm in size, filling the Rosenmüller fossa in the right nasopharynx and showing extension to the right lateral wall in the inferior oropharynx (Figures 2, 3).

Under local anesthetic and 0° endoscopy guidance, multiple punch biopsies were taken from the mass, which was hard, smooth-surfaced, and originated in the right posterolateral nasopharynx. In the histopathological examination, reactive changes were determined. As the pathology and the clinical findings were not compatible, multiple punch biopsies under sedation were taken for a second time from the hard mass originating from the right Rosenmüller fossa. In the histopathology report, fibroblast proliferation was observed in the lymphatic-rich tissue samples and there was no necrosis or mitotic activity. Later, under general anesthesia and 0° endoscopy guidance, the mass in the right posterolateral nasopharynx was almost completely excised, protecting the surrounding tissues. Bleeding control was achieved with bipolar cauterization. The patient was discharged on postoperative Day 1 and was monitored with endoscopy at monthly intervals.

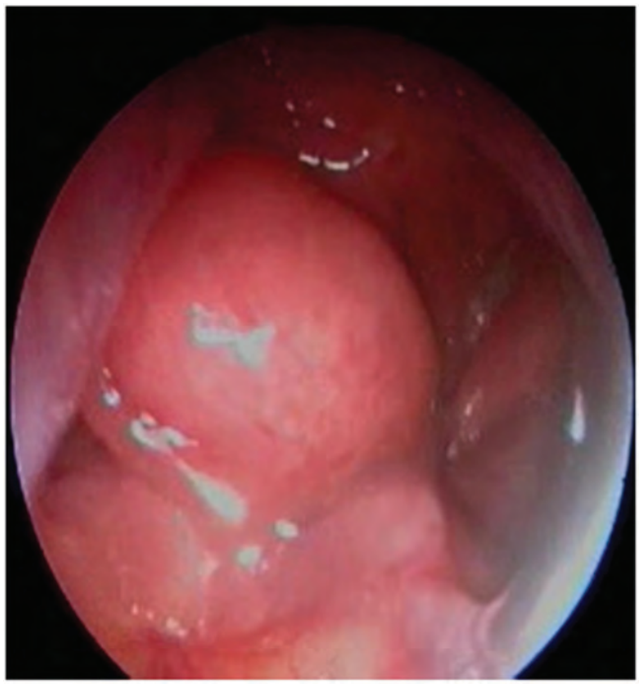

Figure 1. Endoscopic view of the smooth-surfaced mass filling the right Rosenmüller fossa in the nasopharynx

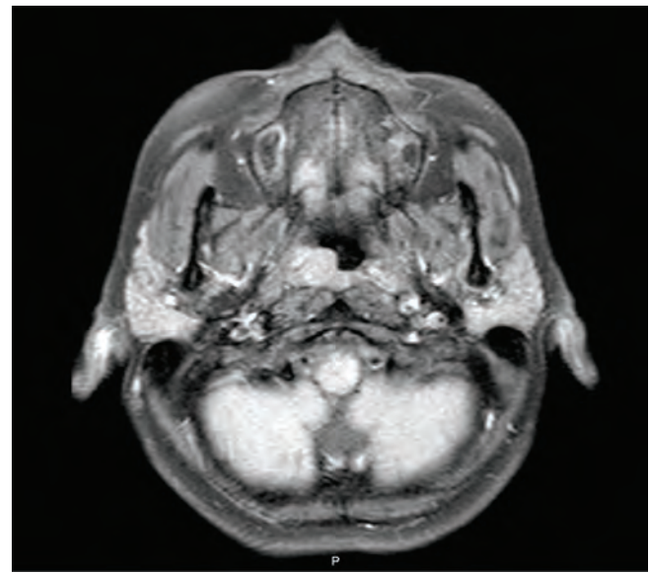

Figure 2. T1 axial slice MR image of the mass filling the right Rosenmüller fossa in the nasopharynx

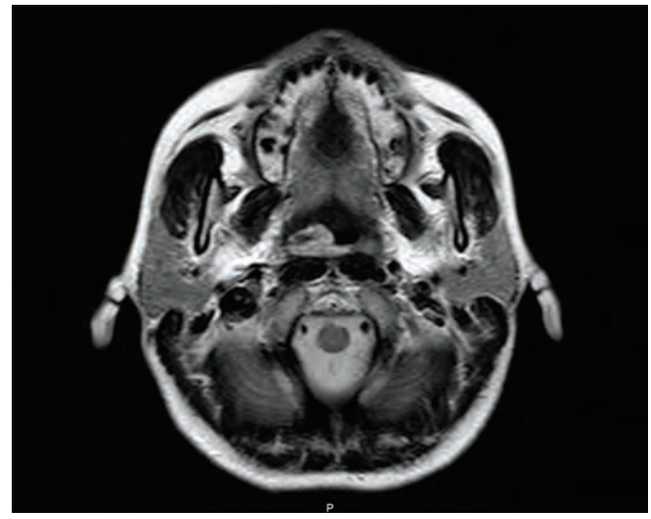

Figure 3 . T2 axial slice MR image of the mass extending to the oropharynx

In the histopathological examination of the excised specimen, an increase was observed in fibrin on the surface and in the lymphatic and vascular structures below the epithelium. The lymphatic structures were observed with CD31 staining and the vascular structures with CD34 (Figures 4, 5, 6). The findings were consistent with lymphangioma.

Sclerotherapy was recommended for the remaining residual tissue, but the patient refused any further treatment as she had

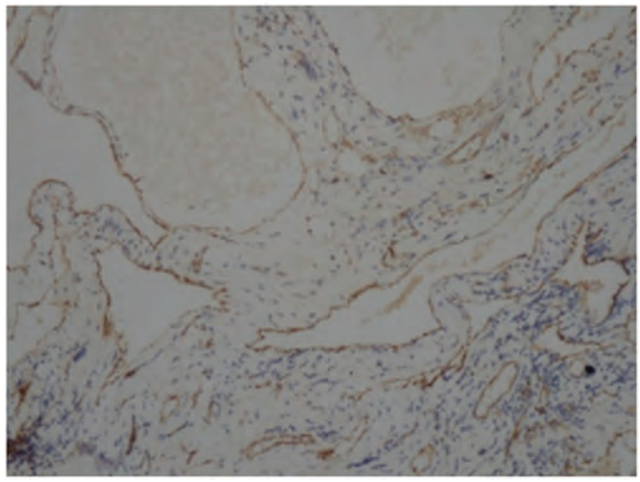

Figure 4. Lymphatic structures stained with CD31 (x100)

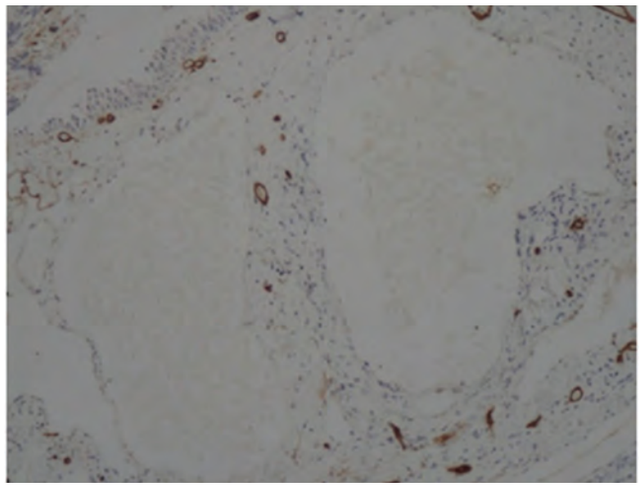

Figure 5. Vascular structures stained with CD34 (x100)

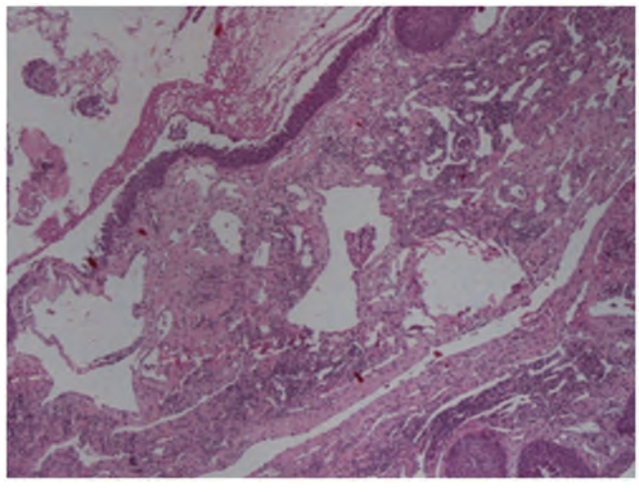

Figure 6. Staining with hematoxylin and eosin (x40)

no complaints. The patient was monitored postoperatively with monthly endoscopic examinations. No growth was determined in the mass which had been reduced in the right lateral nasopharynx (Figure 7). The patient complaints of fullness in the ear and snoring abated. On the nasopharyngeal MRI taken 6 months after the reduction of the mass, an increase was determined in asymmetrical thickness in the Rosenmüller fossa in the right nasopharynx extending to the right lateral wall. The patient had no complaints during the 1-year clinical follow-up period, and no further growth was observed in the mass.

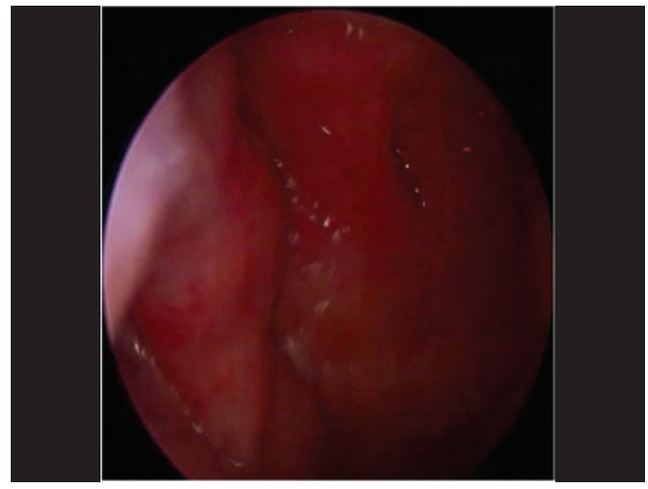

Figure 7. Endoscopic view of the nasopharynx at the 1-year follow-up examination

Discussion

Lymphangioma are benign hamartomatous malformations of the lymphoid vessels. These malformations may be congenital or develop later. However, 90% are seen under the age of 2 years and only extremely rarely in adults. Those that are congenital often accompany chromosomal anomalies such as Turner syndrome and diagnosis is made in the antenatal period with fetal ultrasound. Here we report a case of a 40-year-old with a nasopharyngeal lymphangioma. Lymphangioma that develop later occur as a result of trauma, inflammation, or lymphatic obstruction. There was no trauma story and no chromosomal abnormalities in our present case. Although lymphangioma may occur in any area of the body, they are most frequently seen in the head and neck area. However, isolated lymphangioma with nasopharyngeal location are extremely rare, with two cases published in the Russian literature in 1966 and 1969 [3,4] and two cases in the English literature in 2013 and 2014 [1,4]. To the best of our knowledge, the case presented in this paper is only the 5th reported case in the literature of isolated nasopharyngeal lymphangioma. The previous two most recent cases were males and the current case was a female [1,4].

The symptoms of oral cavity and larynx lymphangioma are dysphagia, dyspnea, the feeling of a foreign body in the throat, throat pain, and frequent attacks of tonsillitis. The cases reported in the literature of isolated nasopharyngeal location presented at ENT polyclinics with complaints of nasal obstruction and irritation in the throat. As in the current case, nasopharyngeal lymphangioma may appear with findings of Eustachian dysfunction. Unlike previous cases in the literature associated with obstruction, the primary complaint on presentation of the current case was snoring.

Endoscopic visualization of the mass is sufficient for advanced testing. Radiological imaging is helpful in the diagnosis of lymphangioma. MR can differentiate the mass from surrounding tissue and can provide information about the size of the mass [5]. The size of the mass in this case was similar to those in previously-reported cases. In the differential diagnosis, nasopharyngeal carcinoma, nasopharyngeal angiofibroma, nasopharyngeal cystic lesions, and other benign masses of the nasopharynx should be considered. In the current case, a polypoid mass was observed filling the right Rosenmüller fossa; this

requires histopathological evaluation for a definitive diagnosis. In the current case, diagnosis could be made only with almost total excision of the mass under general anesthesia, following two previous punch biopsies.

Most lymphangiomas represent malformations rather than true neoplasms and are thought to result from failure of the lymphatic system to communicate with the venous system. Microscopically, lymphangioma consist of large lymphatic channels growing in loose connective tissue. A few disorganized bundles of smooth muscle can be present in the wall of the larger channels. Focal areas of papillary endothelial proliferation similar to those described in blood vessels are sometimes found [6]. Large collections of lymphocytes may be present in the stroma and cause mistakes in interpretation. Lymphangioma almost never become malignant and are curable by excision [7]. In the current case, fibrin was observed on the surface along with an increase in lymphatic and vascular structures below the epithelium; the lymphatic structures were observed with CD31 staining and the vascular structures with CD34.

Lymphangioma are progressive and do not spontaneously regress like hemangioma. Therefore, the disease must be treated. Various methods are used in the treatment of lymphangioma such as surgical excision, reducing the mass with lasers, sclerotherapy, and corticosteroids [5]. As bleeding and edema are less in the laser method, it has recently become a preferred method. Various sclerosing agents such as sodium morrhuate, dextrose, tetracycline, doxycycline, bleomycin, ethibloc (alcohol), and OK432 (lyophilized incubation mixture of group A Streptococcus pyogenes of human origin) are effective by inducing fibrosis [8]. Surgery is the basic treatment option for most lymphangioma. However, as lymphangioma do not have a capsule, most clinicians do not recommend surgery for non-growing lymphangioma because of the difficulties in protecting adjacent vital structures in surgical intervention and high recurrence rates. In the current case, surgery was recommended because the mass was narrowing the passage and because of the patient's complaints. With a transnasal approach with 0° endoscopy protecting the surrounding structures, the mass in the nasopharynx was almost totally excised. In the 1-year follow-up period, there were no clinical complaints and no significant mass was observed in the endoscopic view of the nasopharynx. As the longest follow-up period for recurrence in the literature is 18 months, long-term follow-up results have not yet been reported [1].

In conclusion, although lymphangioma in the nasopharynx are rare, they do occur. This must certainly be kept in mind in the differential diagnosis of patients with symptoms of nasal obstruction and patients seen to have a mass lesion in the nasopharynx. In treatment, a surgical method that protects adjacent vital structures must be used. As there is a risk of recurrence of lymphangioma, patients must be regularly monitored with endoscopy and radiology.

Competing interests

The authors declare that they have no competing interests.

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Bone SPECT/CT in the Evaluation of Soft Tissue Calcifications in a Patient with Dermatomyositis

Dermatomyozitli Bir Hastada Yumuşak Doku Kalsifikasyonlarının Değerlendirilmesinde Kemik SPECT/BT

Bone SPECT/CT In Dermatomyositis

Pelin Arıcan, Seniha Naldöken, Bernatekin Okudan
Nuclear Medicine Department, Ankara Numune Education and Research Hospital, Ankara, Turkey

28. Ulusal Nükleer Tıp Kongresi, İzmir, Türkiye (2016). Poster presentation

Öz

Dermatomiyozit, nadir bir otoimmün hastalıktır. Dermatomiyozitli hastalarda yumuşak dokulardaki kalsiyum fosfat komplekslerinin birikimi sık görülür. Tüm vücut kemik sintigrafisi (TVKS) yumuşak doku kalsifikasyonunu değerlendirmede kullanılabilir. Yumuşak doku kalsifikasyonunun belirlenmesinde, tedavi ve hastalık aktivitesinin planlanmasında ve izlenmesinde önemli bir rol oynamaktadır. Burada, yumuşak doku kalsifikasyonlarının lokalizasyonu ve yaygınlığını değerlendirmek üzere kemik sintigrafisi için gönderilen dermatomyozitli bir hastanın TVKS ve SPECT/BT bulgularını sunuyoruz.

Anahtar Kelimeler

Dermatomyozit; Sintigrafi; Tek Foton Emisyonlu Bilgisayarlı Tomografi SPECT/BT

Abstract

Dermatomyositis is a rare autoimmune disorder. The accumulation of calcium-phosphate complexes in soft tissues is frequent in patients with dermatomyositis. Whole body bone scintigraphy (WBBS) can be used in assessing the soft tissue calcification. It plays an important role in the detection of soft tissue calcification, planning and monitoring of treatment and disease activity. Here in, we present the findings of WBBS and SPECT/CT in a patient with dermatomyositis who is referred for bone scintigraphy to assessment of localization and extent of soft tissue calcifications.

Keywords

Dermatomyositis; Scintigraphy; Single Photon Emission Computerized Tomography; SPECT/CT

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Corresponding Author: Pelin Arıcan, Nuclear Medicine Department, Ankara Numune Education and Research Hospital, 06300, Altındağ, Ankara, Turkey.
T.: +90 3125084877 F.: +90 3123126876 E-Mail: psarican@yahoo.com

Introduction

Dermatomyositis is a rare connective tissue disorder characterized by muscle inflammation, symmetric skeletal muscle weakness, and skin rashes. It is occasionally associated with systemic involvement such as dysphasia, polyarthritis and interstitial pulmonary disease. Diagnosis is based on clinical examination, high muscle enzymes and biopsy. The calcifications in soft tissue are frequently seen in patients. Corticosteroids and immunosuppressive agents are used in treatment. The excision of painful calcification areas may be required in some patients [1]. Whole body bone scintigraphy (WBBS) plays an important role in the detection of soft tissue calcification, planning and monitoring of treatment and disease activity [1,2]. Correlation of anatomical and functional findings with Single Photon Emission Computed Tomography/ Computed Tomography (SPECT/CT) is increased diagnostic accuracy of WBBS [3].

We present the findings of WBBS and SPECT/CT in a patient with dermatomyositis who is referred to assessment of localization and extent of soft tissue calcifications.

Case Report

A 77-year-old woman who had progressive muscle weakness, pain and erythematous plaques on her face had been diagnosed dermatomyositis with high muscle enzyme and muscle biopsy in 2006. Corticosteroid therapy was started and decreased patient's complaints. In 2008, methotrexate treatment was added when her complaints increased. When painful hard wounds around the bilateral gluteal regions were occurred in 2014, colchicine therapy was given for calcinosis cutis. Pamidronate and intravenous immunglobuline were started on the exacerbation of dermatomyositis symptoms. The findings of laboratory revealed creatinin kinasez value of 94 U/L (normal value range: 10-171U/L), alanine transaminase of 14 U/L (normal value range: 3-50 U/L), aspartate transaminase of 30 U/L (normal value range: 4-50 U/L), gamma glutamyl transpeptidase of 17 U/L (normal value range: 6-55 U/L), ALP of 43 U/L (normal value range: 30-120 U/L), C-reactive peptide of <0,2 mg/L (normal value range: 0,2-5 mg/L), romatoid factor of 9 IU/mL (normal value range: 0-14 IU/mL), erythrocyte sedimentation rate of 66 mm/h (normal value range: 0-20 mm/h). The extensive myopathy in the proximal and distal skeletal muscle was found by electromyography. Gluteal ultrasonography and abdominal computed tomography revealed the extensive subcutaneous soft tissue calcifications in the bilateral gluteal region and upper thigh, abdomen and pelvis. Bilateral femur magnetic resonance showed the oedematous and infectious changes from proximal to distal thigh. Lithotripsy was planned for the calcinosis cutis. Bone scintigraphy was performed to assess the prevalence and localization of soft tissue calcifications in the whole body before lithotripsy. WBBS was obtained 3 hours after intravenous injection of 740 MBq Technetium-99m Methylene Diphosphonate (Tc-99m-MDP). After the planar bone scan, SPECT/CT was made from the pelvis which was the most intense of soft tissue involvement. Whole body images showed the heterogeneous radiotracer uptake within soft tissue around the right elbow, the bilateral iliac crest and gluteal regions, left knee and right cruris (Fig.1). Due to the excess of soft tissue involvement in the pelvis, the contours of the bone could not be seen in somewhere. In the SPECT/CT study, radiotracer uptakes of pelvic region were seen in the lateral wall of the lower abdomen, lateral of the bilateral iliac crest, both gluteal regions, coxigius and upper inner of both thighs correspond to the irregular diffuse calcification areas within soft tissue (Fig.2). Lithotripsy was not appropriate because of the wide areas of calcinosis cutis according to the findings of whole body scan. The surgical treatment was decided for calcinosis cutis.

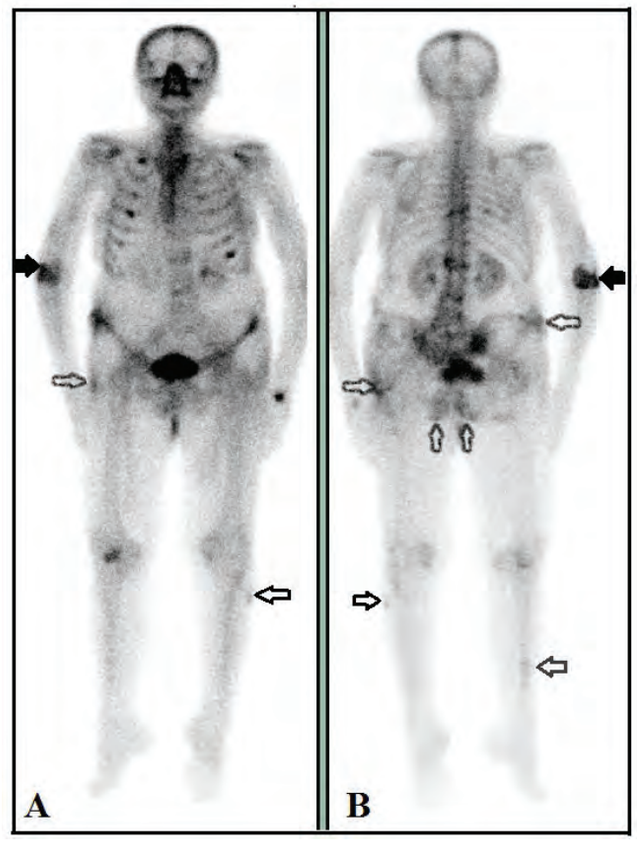

Figure 1: Anterior (A) posterior whole body scintigraphy (B) planar images showed the soft tissue radiotracer uptake right elbow (black arrow), around the bilateral iliac crest and gluteal regions (thin arrows), left knee and right distal tibia (thick arrows).

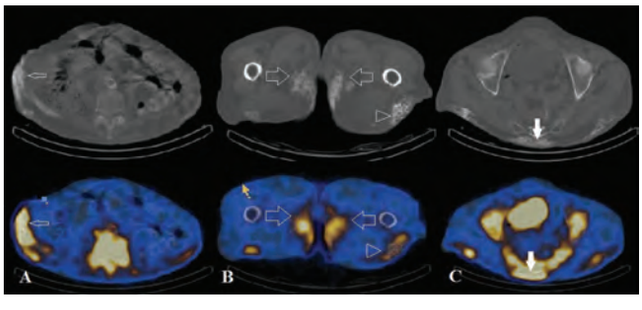

Figure 2: After the whole body scintigraphy, Single emission photon computed tomography was performed to the region of abdomen and pelvis. Axial Computed Tomography and fusion images showed the subcutaneous soft tissue calcification areas associated with nonhomogeneous increased osteoblastic activity in the lower abdominal wall (thin arrows) (A), bilateral proximal thighs (thick arrows), bilateral gluteal regions (triangular) (B), and around the coccygeus (white arrows) (C).

Discussion

Dermatomyositis is chronic inflammatory disease of skeletal muscle. It is thought that the cause of inflammation in the muscles belongs to autoimmune response. As a result of autoimmune damage in the muscles, calcium complexes in the

muscles and soft tissue accumulate in patient with dermatomyositis [1]. Tc-99m phosphonate used in bone scintigraphy may accumulate extra osseous tissue in some conditions such metastatic calcifications, McArdle Syndrome, rhabtomyolisis, myositis ossification, and polydermatomyositis [4-6]. Bone scan can be performed to assess the soft tissue calcifications in the dermatomyositis. There is a correlation between the intensity of radiotracer uptake and disease activity. Therefore bone scintigraphy can be used for management of the patients and evaluation of response to the therapy [4,5,7]. Also all soft tissue involvement from head to feet is evaluated simultaneously with WBBS. Whole-body imaging of patients with conventional imaging methods may not be possible. The addition of SPECT/CT to planar images identifies the accurate anatomic location and extent of radiotracer uptake in bone scintigraphy. Also it may detect more lesion than planar bone scan. It is increased the diagnostic accuracy and specificity of bone scintigraphy [3]. In this case, bone scintigraphy findings were useful for detecting extensive calcification in the whole body. WBBS showed unexpected soft tissue calcinosis in the elbow, left knee and right 1/3 distal tibia without conformity of diagnostic work up. SPECT/CT detected to precise anatomic localization of calcifications and additional calcification areas as bilateral proximal thighs which could not be seen with WBBS [6]. According to the findings of WBBS and SPECT/CT, the planning of therapy was changed. Lithotripsy procedure was abandoned because of extensive calcification areas. Excision of painful calcification regions was decided.

As a conclusion, WBBS and SPECT/CT are useful assessing the soft tissue involvement and calcinosis cutis in patients with dermatomyositis. Bone scintigraphy shows the prevalence of the soft tissue calcinosis as it allows whole body imaging and it changes the patient's treatment.

Competing interests

The authors declare that they have no competing interests.

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The Diagnosis and Response to Treatment of an Extrapulmonary Sarcoidosis on F^{18}-FDG PET/CT: A Case Report

Ekstrapulmoner Sarkoidozda Tanı ve Tedavide F^{18} FDG PET/BT: Bir Olgu

Hatice Sınav Uslu[1], Halil İbrahim Yakar[2], Asiye Kanbay[2], Mehmet Tarık Tatoğlu[1], Serkan Güngör[1]
[1]Department of Nuclear Medicine, [2]Department of Pulmonology, Faculty of Medicine, Istanbul Medeniyet University, Istanbul, Turkey

Özet

Altmış dokuz yaşında kadın hasta, bacaklarda deri döküntüleri ile hastanemize başvurdu. Cilt biyopsisinde eritema nodosum tanısı konuldu. Bronkoskopik biyopsi, kronik non-nekrotizan granülomatöz enflamasyon olarak raporlandı. FDG PET/BT'de hipermetabolik multipl lenfadenopati, pulmoner nodüller ve masif splenomegali görüldü. Dalak rüptürü riski nedeniyle hastaya oral kortikosteroid tedavisi verildi. Tedavi sonrası kontrol FDG PET/BT' de dalakta ve multipl lenfadenopatilerde tam gerileme gözlemlendi. FDG PET/BT görüntüleme akciğer dışı sarkoidoz tedavi kararı verilmesinde ve tedaviye yanıtın takibinde yararlıdır.

Anahtar Kelimeler

FDG PET/BT; Ekstrapulmoner Sarkoidozis; Tanı; Tedavi

Abstract

A 67-year-old female patient was admitted to our hospital with skin eruptions on her legs. The skin biopsy verified a diagnosis of erythema nodosum histopathologically. The bronchoscophic biopsy was reported as chronic non-necrotizing granulomatous inflammation. FDG PET/CT revealed hypermetabolic multiple lymphadenopathy, pulmonary nodules and giant splenomegaly. The patient was treated with oral corticosteroid for the risk of splenic rupture. A complete regression was observed in the spleen and multiple lymphadenopathy. FDG PET-CT imaging may be useful to establish the treatment decision and response to treatment of extrapulmonary sarcoidosis.

Keywords

FDG PET/CT; Extrapulmonary Sarcoidosis; Diagnosis; Treatment

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Corresponding Author: Halil Ibrahim Yakar, Department of Pulmonology, Faculty of Medicine, Istanbul Medeniyet University, 34722, Istanbul, Turkey.
T.: +90 2165664000 F.: +90 2165666614 E-Mail: halil_yakar@hotmail.com

Introduction

Sarcoidosis is a systemic disease that can involve all organ systems [1]. It is seen especially in the lungs and lymph nodes but it may also be seen as extrapulmonary sarcoidosis (ES). For diagnosis of ES, radiological methods are used such as scintigraphy, USG and MRI. In recent years, 18-fluorodeoxyglucose positron-emission tomography/computer tomography (FDG PET/CT) can also be used for diagnosis of ES. We present a rare case of sarcoidosis with multisystem involvement identified on FDG PET/CT.

Case Report

A 67year-old female patient was admitted to our hospital with skin eruptions on her legs that had not responded to treatment by local steroid. The skin biopsy verified a diagnosis of erythema nodosum histopathologically. The blood Angiotension Converting Enzyme (ACE) level was found to be 84 U/l. Transbronchial Needle Aspiration with fiberoptic bronchoscopy revealed chronic non-necrotizing granulomatous inflammation. The FDG-PET/CT detected multiple hypermetabolic bilateral chest lymphadenopathy, a butterfly-shaped distribution pattern which is typically seen in patients with sarcoidosis mediastinal lymph nodes, multiple abdominal, inguinal lymph nodes, and hypermethabolic splenomegaly (FIGURE 1-2-3-4). Therefore, the patient was treated with oral corticosteroid (OCS) throughout one year. A complete regression was observed in the spleen and multiple lymphadenopathy after 3 months initiationof treatment (FIGURE 5).

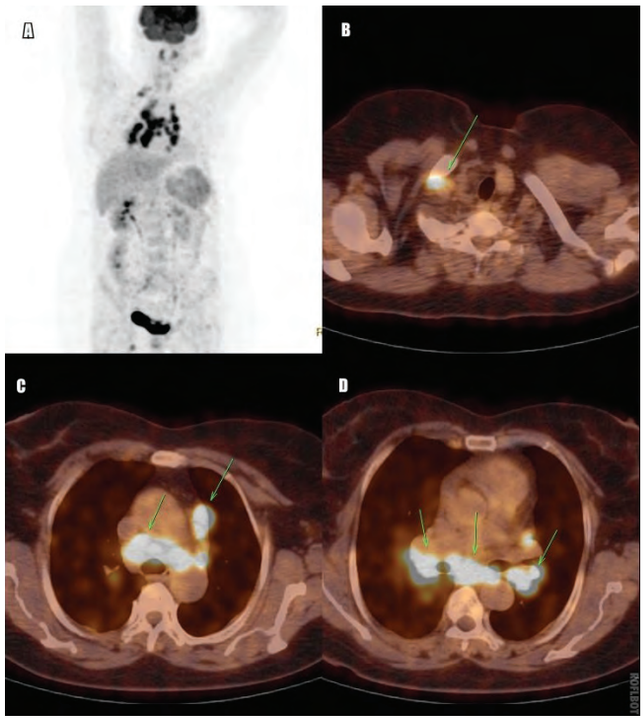

Figure 1. MIP image. FDG PET/CT axial images revealed intensely hypermetabolic bilateral chest lymphadenopathy in a butterfly-shaped distribution pattern, which was typically seen in patients with sarcoidosis(A). Supraclavicular and infraclavicular hypermetabolic multiple LAPs which the largest was 16 mm sized (SUDmax: 8.5) (B). Intensely hypermetabolic LAPs which the biggest was 27 mm diameter (SUDmax: 14.0) were observed in the mediastinal prevascular area, right upper and lower paratracheal, aortopulmonary, carinal, subcarinal, paraesophageal and bilateral hilar areas (C-D).

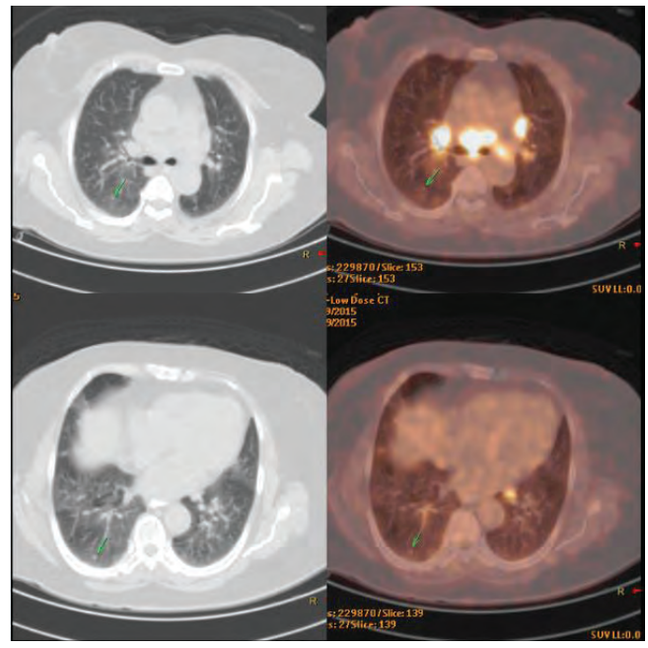

Figure 2. In the right lower lobe superior and posterobasal segments were seen mild hypermetabolic nodular densities which the largest was 9 mm diameter (SUDmax: 3.8).

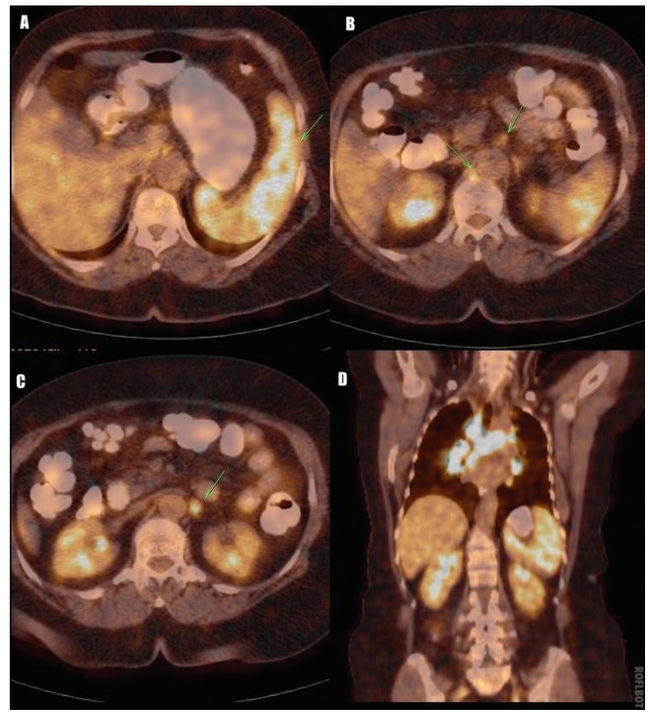

Figure 3. The size of the spleen and FDG uptake were increased (spleen SUDmax: 6.5, liver SUDmax: 3.9) (A). In right retrocrural and left paraaortic area were seen hypermethabolic lymph nodes (SUDmax: 6.2)(B-C) In coronal image was seen hypermetabolic mediastinal lymph nodes and hypermetabolic splenomegaly(D).

Discussion

Sarcoidosis may be a life-threatening disease due to involvement of all organ systems to a varying extent and degree. The lungs and lymph nodes are the most frequently involved organs with a frequency of 90% and 30%, respectively [2]. Less frequently, the skin, liver, and spleen may be involved. Skin involvement of sarcoidosis has been reported between 10-30% [3-4]. Liver involvement is 5-15%. Spleen involvement ranges from 1% to 40% [5-6]. Spleen, liver and skin involvement, as found in this case, is an extremely rare combination in the literature.

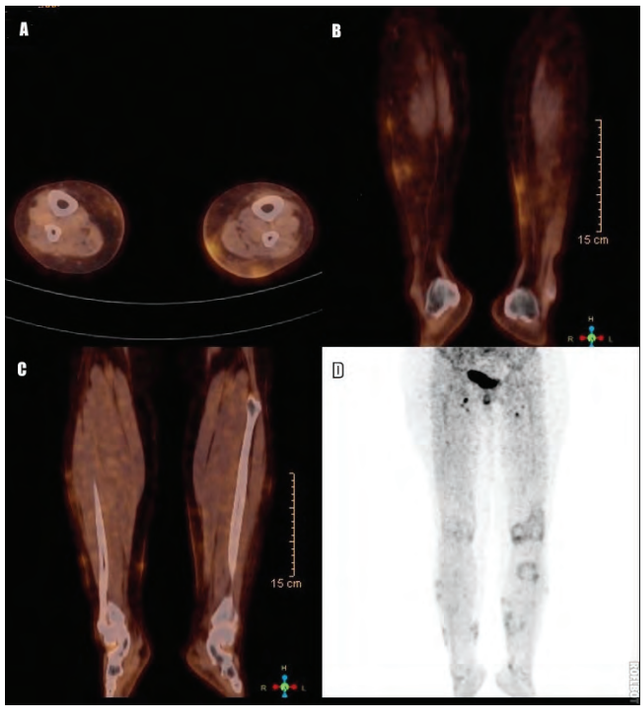

Figure 4. Common heterogen mild FDG uptake was seen in skin of the left medial and in the distal part of both lower extremities (SUDmax: 3.7)(A,B,C,D).

The use of gallium-67 scintigraphy is prevalent in the diagnosis of ES. And an alternative tracer, 68Ga-DOTA-1-Nal3-Octreotide (68Ga-DOTANOC) binds to somatostatin receptors on inflammatory cells in sarcoid granulomas. However, its use has gradually decreased. FDG PET/CT imaging has undergone important advances in recent years and has a higher sensitivity than gallium scintigraphy [7]. FDG PET/CT has a great advantage in the detection of inflammatory, active granulomatous disease in patients with ES [8].

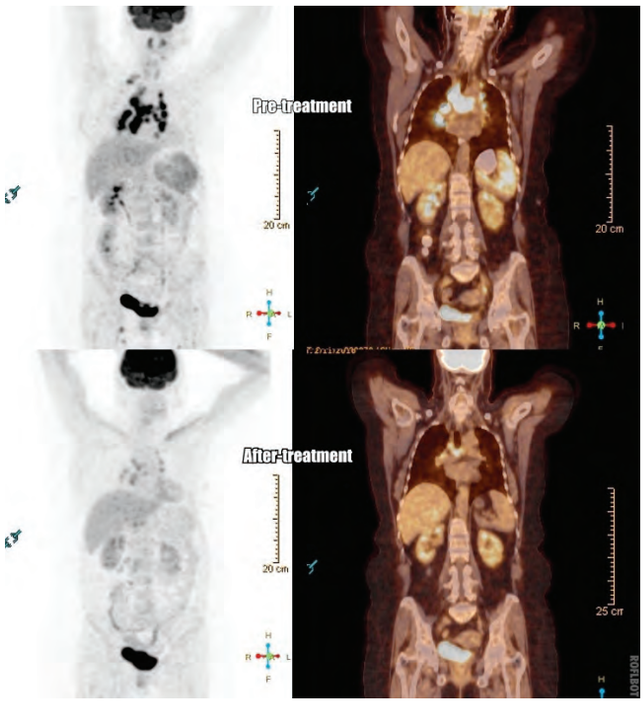

Figure 5. Supra-infraclavicular, mediastinal, intraabdominal hypermetabolic LAPs were observed significant metabolic and morphologic regression on PET-CT images three months after initializing of treatment. In the spleen was observed complete regression findings as metabolic and morphological.

In sarcoidosis, the risk of rupture due to massive splenic involvement is high and splenectomy is required if treatment is not given. Our case also had massive spleen involvement of sarcoidosis as diagnosed via FDG PET CT. However, we observed complete regression of metabolic and morphological changes in the spleen after treatment. So we claim that our case is important to highlight the use of FDG PET/CT in the diagnosis and response to treatment of ES.

Competing interests

The authors declare that they have no competing interests.

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Leriche Syndrome in the Emergency Department: Two Case Reports

Acil Serviste Leriche Sendromu: İki Olgu Raporu

Leriche Syndrome

Mustafa Yilmaz[1], Mehtap Gurger[1], Metin Atescelik[1], Mehmet Cagri Goktekın[1], Ihsan Yigit[2]
[1]Department of Emergency Medicine, Firat University School of Medicine, Elazig,
[2]Department of Emergency Medicine, Basaksehir State Hospital, Istanbul, Turkey

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Özet

Aortik bifürkasyonun oklüzyonu sonucu meydana gelen Leriche sendromu, cerrahi olarak tedavi edilen, ciddi morbidite ve mortaliteye neden olan nadir görülen periferik vasküler hastalıklardandır. Tek yada iki taraflı alt ekstremitede kladicasyo, femoral nabızların azalması yâda yokluğu ile karakterizedir. Alt ekstremitelerde ağrı ya da güçsüzlük şikâyeti olan hastalarda ayırıcı tanıda düşülmelidir.

Anahtar Kelimeler

Acil Servis; Leriche Sendromu; Trombozis

Abstract

Leriche syndrome, resulting in occlusion of the aortic bifurcation, is a rarely seen peripheral vascular disease that is treated surgically and causes severe morbidity and mortality. It is characterized by claudication in one or both lower extremities and by absent or decreased femoral pulses. It should be considered in the differential diagnosis in patients with complaints of pain or weakness in the lower extremities.

Keywords

Emergency Service; Leriche Syndrome; Thrombosis

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Corresponding Author: Mustafa Yilmaz, Department of Emergency Medicine, Firat University Faculty of Medicine, 23200, Elazig, Turkey.
T.: +90 4242370000/1443 GSM: +905057700734 E-Mail: drmylmz@hotmail.com

Introduction

Leriche syndrome (LS), first described in 1923, is a clinical condition with a high mortality, which usually occurs due to thrombosis at the infrarenal aorta or bilateral iliac artery bifurcations on an atherosclerotic basis [1].The thrombus is usually at the level of the aortic bifurcation and can reach the level of the renal arteries over time. The mortality and morbidity rates of LS have been reported to be between 4.5-5.0% and 18-20%, respectively [2-6]. Two forms have been described, namely acute and chronic [7]. In the acute form, ischemic symptoms as a result of acute occlusion of peripheral arteries include leg pain, pulselessness, paresthesia, decreased temperature, pallor, and paralysis due to thrombosis in the aorta and iliac arteries. Symptoms and signs of intestinal ischemia or renal failure due to impaired blood flow in intestinal and renal arteries may develop. Leriche syndrome often occurs in males over 50 years of age, and it characteristically causes claudication in one or both lower extremities, absent or decreased femoral pulses, and erectile dysfunction in males [1,2]. In the literature, atypical findings such as paraplegia, urinary incontinence, flank pain, and shortness of breath have been reported according to the occluded level of the abdominal aorta [8-10]. The main treatment is surgery in LS, but angioplasty and endovascular stenting are the other treatment options in cases of focal involvement [11]. Abdominal aortic dissection, vertebral degenerative diseases, mechanical pain due to disc herniation, neuropathy, and Guillain-Barre syndrome should be considered in the differential diagnosis.

In this article, we aimed to present two LS cases that presented with symptoms of paresis, pain, and bruising in both legs.

Case Report 1

A 77-year-old female patient was admitted to the emergency room with complaints of bruising and pain in the foot. Her medical history included hypertension, type 2 diabetes mellitus, and an ischemic stroke 15 days before., It was learned that she was taking her low molecular weight heparin, antihypertensive, and oral antidiabetic drugs regularly. The patient had pain, bruising, and coldness in both legs that had started approximately 10 hours before and that had increased progressively. Her vital signs were as follows: blood pressure=150/100 mmHg, pulse=88 beats/min, respiratory rate=18/min, temperature=36 °C, and oxygen saturation=97%. Physical examination revealed pulselessness in the popliteal-dorsalis pedis and posterior tibial arteries in both lower extremities, and coldness and ecchymotic areas on both lower extremities. Other systemic examinations were normal except for the sequel flat left nasolabial fold from a previous ischemic stroke. Laboratory tests were as follows: WBC=10.4x103/mm3, Hg=17.1 g/dL, Hct=55.3%, APTT=15.7 sec, INR=1.003, Troponin=0.48 ng/mL, creatinine=1.50 mg/dL, CK=233 U/L, and CKMB=105.8 U/L. Both lower extremity arterial color Doppler examinations showed no blood flow to the main femoral arteries or distally. The occlusion was found to extend to the upper parts of the main femoral artery. Abdominal CT angiography revealed that there was no contrast filling from the proximal part of the right main internal iliac artery and that there was blood flow in the left main internal iliac artery, but there was no contrast filling after the external iliac artery due to thrombosis (Figure 1, 2). The patient was diagnosed as having LS, and underwent emergent embolectomy by the Cardiovascular Surgery Clinic and was discharged one week later.

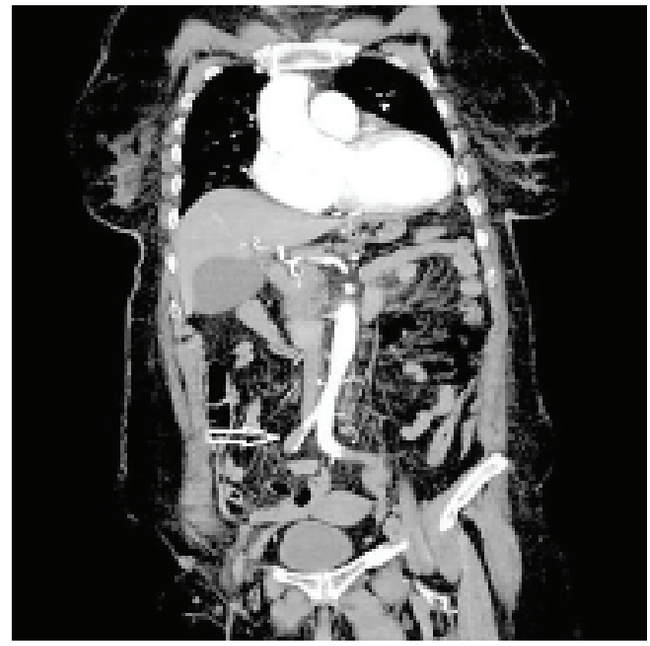

Figure 1. The appearance of total occlusion of main iliac arteries (Case 1)

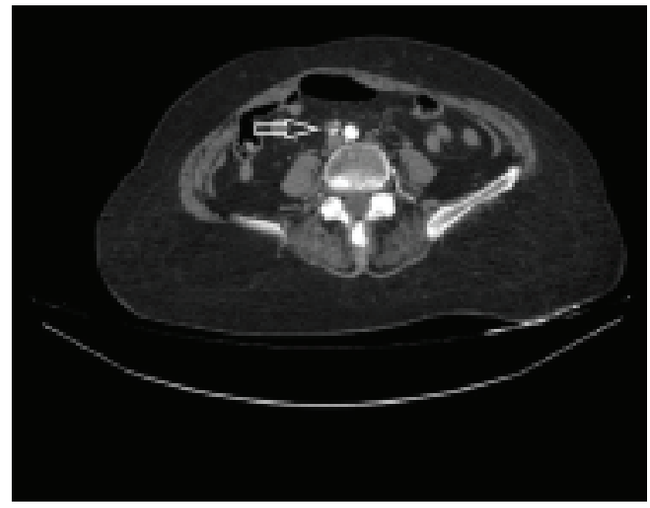

Figure 2. The appearance of total occlusion of the right main iliac artery (Case 1)

Case Report 2

A 72-year-old male patient was admitted to the emergency department with a complaint of weakness in both lower extremities. His medical history included lumbar disc herniation, as well as a surgery due to bladder tumor. He had mild difficulty in walking and leg pain during the previous two days, but was admitted to the emergency department due to acute plegia and anesthesia in both legs for the previous 1-hour. His vital signs were as follows: blood pressure=140/90 mmHg, pulse=104 beats/min, respiratory rate=22/min, temperature=36 °C, and oxygen saturation=96%. On physical examination, bilateral lower extremity was paraplegic and deep tendon reflexes were negative. Although the pulses were weak in the femoral, popliteal-dorsalis pedis, and posterior tibial arteries, the lower extremities were cold and pale. Laboratory tests were as follows: WBC=11.3x103/mm3, Hg=13.6 g/dL, Hct=40.8%,

Troponin=<0.01 ng/mL, creatinine=0.88 mg/dL, CK=35 U/L, and CKMB=<2.0 U/L. An abdominal CT angiography revealed a thrombus that led to occlusion in the arterial lumen extending from the infrarenal part of the abdominal aorta to the bilateral main femoral arteries (Figure 3 and 4). The patient was diagnosed as having LS, and underwent emergent surgery. The patient died one day after the operation.

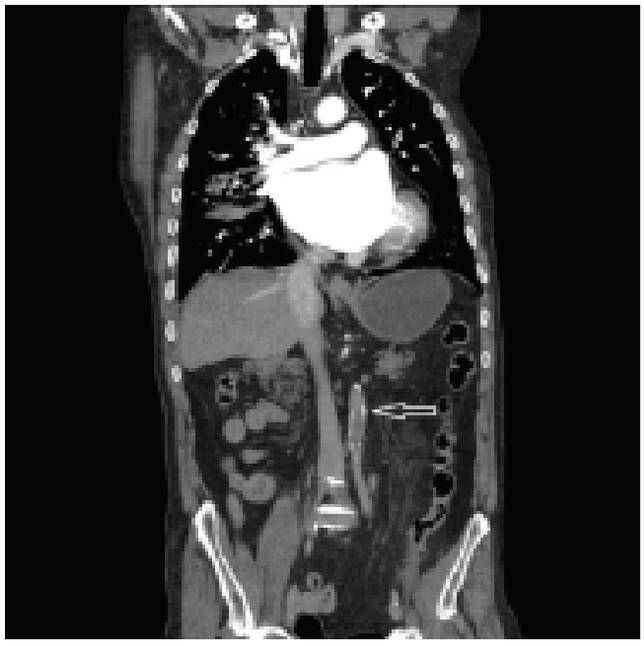

Figure 3. The appearance of total occlusion of the abdominal aorta at the infrarenal level (Case 2)

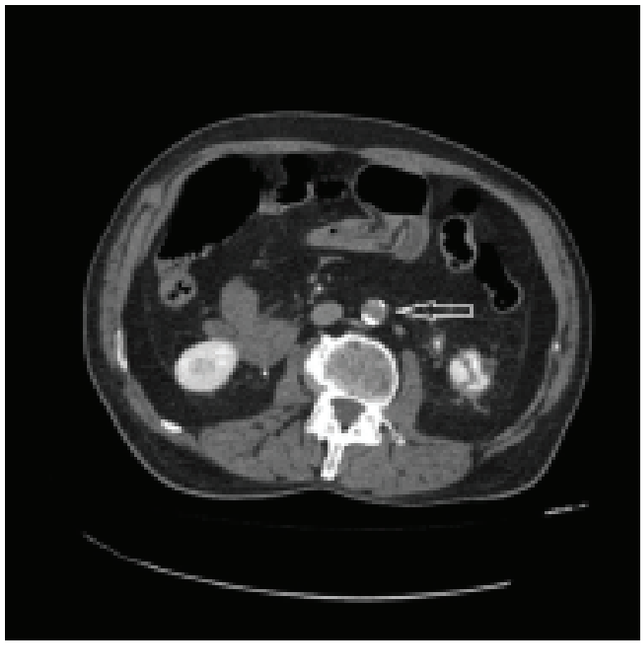

Figure 4. The appearance of total occlusion of the abdominal aorta at the infrarenal level (Case 2)

Discussion

Leriche syndrome is an acute or chronic urgent condition with an atherosclerosis background that frequently requires cardiovascular surgery [7]. Symptoms of acute total occlusion are sudden and severe. The higher mortality in acute occlusions suggests that this disease should be diagnosed early and that treatment should be initiated without delay. Findings related to acute occlusion of peripheral arteries such as pain, pulselessness, paresthesia, coldness, pallor, and paralysis are observed in the limbs due to thrombosis in the aorta and iliac arteries. Depending on the level of occlusion, symptoms such as abdominal pain and renal failure may also develop due to impaired blood flow in intestinal and renal arteries. Flask paraplegia as a result of anterior spinal cord ischemia in the early period of acute aortic thrombosis will rarely be the first finding. It may cause the patients to not recognize the pain, one of the classical symptoms of arterial occlusion in patients, thus causing a delay in treatment [12]. Findings related to the dysfunction of affected organs can occur in chronic occlusion. Bayır et al. [11] reported chronic renal failure in a 50-year-old female patient with loss of strength and pain in both legs, as well as abdominal pain. While there was pain, paleness, and coldness that suggested acute obstruction, as in our first case, her presentation to the emergency department was delayed because the numbness in her legs increased gradually over a period of days and because it was thought that the complaints were caused by previously existing lumbar disc herniation.

Initiation of surgical and medical treatment in the early period after diagnosis of Leriche syndrome in the emergency department will prevent deterioration of the quality of life of the patients by avoiding mortality and prevention of organ loss or loss of extremity. However, diagnosis in the emergency department is often delayed because findings suggest other illnesses or the symptoms are perceived as motor deficits. Late admission in our second case due to the presence of numbness rather than pain in the legs supports findings in the literature that the disease is more mortal in cases of late admission.

Conclusion

Leriche syndrome should be considered in the differential diagnosis of each patient who complains of leg pain or numbness in the emergency department. For this reason, the presence/absence of peripheral arterial blood flow should be evaluated, and patients with suspected pulse filling should be confirmed with color Doppler ultrasonography. Advanced imaging techniques should be used when necessary.

Acknowledgments

Oral consent was obtained from the patients. The authors declare that they have no competing interests.

Competing interests

The authors declare that they have no competing interests.

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Intra-Testicular Leiomyoma: A Case Report

Testisde Yerleşim Gösteren Leiomyom: Olgu Sunumu

Intra-Testicular Leiomyoma

Recep Bedir[1], Rukiye Yılmaz[1], Hüseyin Eren[2]
[1]Department of Pathology, [2]Department of Urology, Recep Tayyip Erdogan University, Medical Faculty, Rize, Turkey

Özet

Leiomyomlar düz kas hücrelerinden köken alan benign tümörlerdir. En sık uterusda görülmek ile birlikte renal pelviste, mesane, spermatik kord, epididim, prostat, skrotum ve glans peniste de görülürler. Ancak tunika albugineada oldukça nadirdir. Biz burada 72 yaşında tunika albugineada yerleşim gösteren bir leiomyom olgusunu sunuyoruz.

Anahtar Kelimeler

Testis; Leiomyom; Radikal Orşiektomi

Abstract

Leiomyomas are benign tumors originating from smooth muscle cells. Though most commonly reported in the uterus, they may also originate from the renal pelvis, urinary bladder, spermatic cord, epididymis, prostate, scrotum, or glans penis. However, location in the tunica albuginea is extremely rare. Herein, we will report a diagnosed leiomyoma located in the tunica albuginea in a 72-year-old patient.

Keywords

Testes; Leiomyoma; Radical Orchiectomy

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Corresponding Author: Recep Bedir, Department of Pathology, Recep Tayyip Erdogan University, Medical Faculty, 53000, Rize, Turkey.
T.: +90 4642130491 F.: +90 4642170364 GSM: +905057331695 E-Mail: bedirrecep@gmail.com

Introduction

Leiomyomas are benign tumors originating from smooth muscle cells. They may arise from the renal pelvis, urinary bladder, spermatic cord, epididymis, prostate, glans penis, or the scrotum in the urinary system. However, leiomyomas located in the testis are extremely rare [1, 2]. To our knowledge, only a few cases have been reported in the literature [1-9]. We herein present a case of intra-testicular leiomyoma.

Case Report

A 72-year-old patient was admitted to our hospital with the complaint of a swelling on his right testis for 1 year. Ultrasonographic evaluation revealed a thick-walled anechoic cyst measuring 32x24 mm in the spermatic canal on superior parts of the right scrotal sac. In the left epididymis, there were also 2 dense cysts having high level echoes with the diameters of 4.5 mm and 6 mm. Para-testicular veins were measured as 2.6 mm at the neutral position and 3.5 mm with Valsalva at the largest points on the left. Ultrasonography (US) of the testes showed a hypoechoic lesion measuring 1 cm in diameter in the lateral parts of the right testis. Eventually, the ultrasound was reported as grade 3 varicocele on the left, a cyst on the right spermatic canal, and 2 epididymis cysts with high density on the left. The patient's serum tumor markers (AFP, β-HCG, LDH) were within normal limits. The patient received radical orchiectomy. In macroscopic evaluation, there was a well-defined solid lesion measuring 0.9x0.6 cm, having a gray-white colored cross-sectional surface, on a side of testis tissue which was 3.5x2 cm in size, just below the tunica albuginea. There was also a cystic structure together with the structures belonging to the epididymis cysts on the right spermatic canal. Microscopic evaluation of the testicular mass revealed a benign tumor characterized by spindle-like cellular proliferations forming crossing bundles. There was not any increased cellularity, tumor necrosis, increased mitotic activity, or pleomorphism determined in the tumor (Figure 1). There were findings of atrophy on the surrounding testis tissue (Figure 2). In immunohistochemical evaluations, the tumor was positively stained with smooth muscle actin (SMA) and desmin (Figure 3). Ki-67 proliferation index of the tumor was low (1-2%). Based on these findings, the case was diagnosed as intra-testicular leiomyoma.

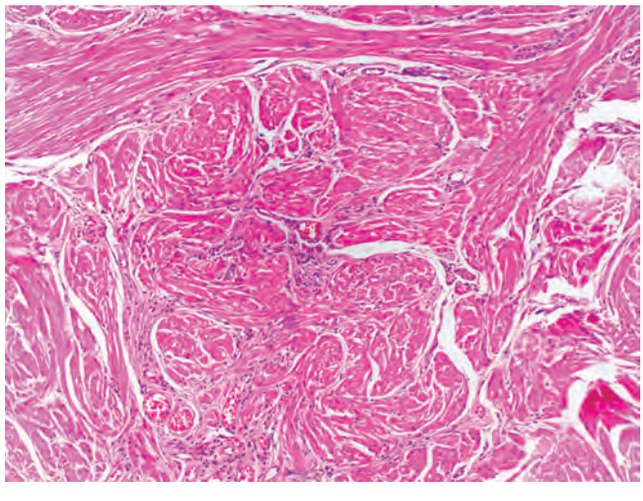

Figure 1. Microscopic examination showed interlacing uniform spindle cells with blunt-ended elongated nuclei (H&E×100).

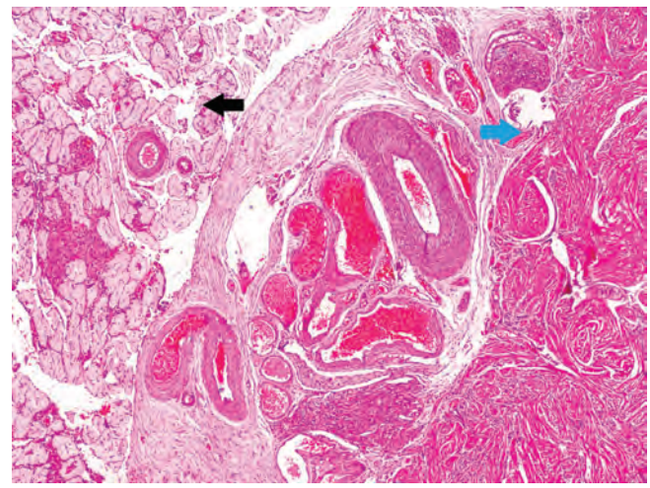

Figure 2. Tumor consists of benign spindle cells (blue arrow) that are near the atrofic testicular tissue (black arrow) (HEx40)

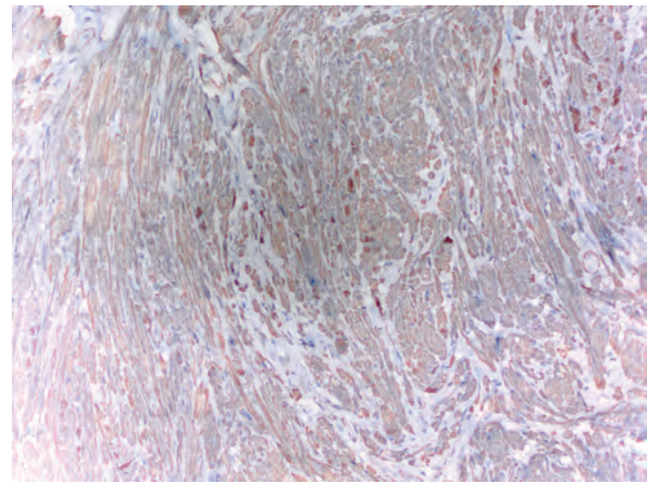

Figure 3. In immunohistochemical staining, the tumor cells were positive with SMA (x200)

Discussion

Most of the solid masses located on the testis are malignant tumors such as germ cell tumors, non-germ cell tumors, lymphoma, and metastatic tumors. Rare benign solid lesions located on the testis include epidermoid cyst, Leyding cell hyperplasia, gonadal stroma originated fibroma, hemangioma, leiomyoma, spontaneous hemorrhages, sarcoidosis and tuberculosis [1]. Leiomyomas located in the testis are extremely rare. Though those tumors may be found in all age groups, they are most commonly reported in the 6th decade of life. Generally, there is a history of painless, slowly growing scrotal swelling for a long time [2-5].

Leiomyomas may arise from any type of smooth muscle cells. The etiology of leiomyomas on the tunica albuginea is controversial. They may originate from the smooth muscle cells of vessels as well as totipotent teratomas [6]. According to another opinion, they originate from the tunica propria of seminiferous tubules or smooth muscle cells of the tunica albuginea [1]. On macroscopic evaluation, they are well-defined, having a whorled cut surface [1-3]. The exact diagnosis is made after histopathological investigations. The differential diagnosis includes inflammatory myofibroblastic tumor (IMT). This tumor is negatively stained with desmin while leiomyomas are positive. In preoperative diagnosis of the tumor, ultrasound may be helpful. In ultrasonographic evaluation leiomyomas are defined as

well-defined, hypoechoic solid lesions; the differential diagnosis includes inflammatory hydrocele, multi-loculated hematocele, and Sertoli cell tumor [2,7-9].

The main treatment method of those tumors is local excision. Radical orchiectomy is unnecessary in general, and should be performed only if the tumor has adhesions to the surrounding testis tissue or if it has a malign appearance. In those conditions, the nature of the surgery should be determined by the aid of the frozen section. However, if frozen section is not available, then since the exact diagnosis usually cannot be made preoperatively, radical orchiectomy becomes the main treatment method. In our case, since frozen section could not be performed, radical orchiectomy was chosen for the treatment.

In conclusion, though intra-testicular leiomyomas are rare, they should be kept in mind in the differential diagnosis of testicular solid masses. However, since most of the testicular solid masses are malignant and preoperative differentiation of benign and malign masses is difficult, most of these patients are treated with radical orchiectomy. For that reason, in patients with the suspicion of benign tumors, it should be kept in mind that with intraoperative consultation (frozen section) these lesions may be treated with only excisional biopsy, thus avoiding unnecessary radical surgeries.

Conflict of Interest

No conflict of interest was declared by the authors.

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Osteonecrosis of the Sesamoid Bones: Two Case Reports

Sesamoid Kemik Osteonekrozu: İki Olgu Sunumu

Osteonecrosis of Sesamoid Bone

Ayhan Aşkın[1], Ece Güvendi[2], Aliye Tosun[1], Özgür Tosun[3]
[1]Department of Physical Medicine and Rehabilitation, Katip Çelebi University, Faculty of Medicine, Izmir,
[2]Physical Medicine and Rehabilitation Clinic, Dumlupınar University Evliya Çelebi Training and Research Hospital, Kütahya,
[3]Department of Radiology, Katip Çelebi University, Faculty of Medicine, Izmir, Turkey

Özet

Sesamoid kemik osteonekrozu, metatarsaljinin nadir bir nedenidir ve tanı konulması zor olabilir. Hastalık, klasik olarak, ayak travma öyküsü olan kadın hastalarda bildirilmektedir. En sık şikayet, ağrıdır. Ağrı, 1. metatarsal kemik üzerine ağırlık verildiğinde, yürüme siklusunun son fazında, halluks ekstansiyonu ile ortaya çıkar. Erken tanıda en faydalı inceleme yöntemi, osteonekrotik sürecin izlenebildiği manyetik rezonans görüntüleme (MRG)'dir. Öncelikli tedavi seçeneği, konservatif yaklaşım olmalıdır. Konservatif tedavilerin, yetersiz kaldığı durumlarda cerrahi tedavi düşünülebilir. Burada başarı ile tedavi ettiğimiz iki olguyu sunmayı amaçladık.

Anahtar Kelimeler

Sesamoid; Metatarsal Ağrı; Osteonekroz

Abstract

Osteonecrosis of the sesamoid bones is an uncommon cause of metatarsalgia and may be difficult to diagnose. The disease is classically reported in female patients with a history of foot trauma. The most frequent complaint is pain. Pain occurs with the extension of the hallux in the final phase of the gait cycle when weight is given on the first metatarsal bone. The most useful examination method in early diagnosis is MRI by which the osteonecrotic process can be monitored. The first treatment option should be a conservative approach. Where conservative treatments prove insufficient, surgical treatment can be considered. In this study, we aimed to present two cases we have successfully treated.

Keywords

Sesamoid; Metatarsal Pain; Osteonecrosis

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Corresponding Author: Ayhan Aşkın, Department of Physical Medicine and Rehabilitation, Katip Celebi Uni. Ataturk Training and Research Hospital, Izmir, Turkey.
T.: +90 2322444444 F.: +90 2322431530 E-Mail: ayhanaskin@hotmail.com

Introduction

Metatarsalgia is a common complaint, frequently encountered in daily practice. Sesamoid bone pathologies are among the causes of metatarsalgia, but are rarely diagnosed [1]. They are small and may seem functionally unimportant, but their pathologies may result in severe pain and dysfunctions as they contribute to the weight-bearing function of the foot and the big toe [2]. These painful conditions may exhibit an acute post-traumatic or insidious onset and may be due to arthritis, infection, or ischemia [3,4]. Physical examination and imaging methods are useful in differential diagnosis. In this case report, our aim was to share our experience with two patients who applied with complaints of metatarsal pain and were diagnosed with osteonecrosis of the sesamoid bone and to review the literature as well.

Case Report 1

A thirty-year-old female patient was admitted to our outpatient clinic with mechanical pain on the plantar face of the right toe that had increased for 10 days. She stated that the pain was worse when weight was given on the big toe and did not decrease with rest. The patient found it hard to step on the right foot due to pain. She did not describe recent trauma. She did not have any additional characteristics in her medical history except a job that required standing for a long time during the day.

In physical examination, the passive and active range of motions of the first metatarsophalangeal (MTF) joint were normal but painful. There was no color change or swelling. The first metatarsal region was oversensitive in palpation. No sensory or motor deficit was found. Laboratory examination was normal. Upon detection of a heterogeneous image consistent with sesamoid bone localization in lateral and anterior-posterior roentgenograms of the foot, a computed tomography (CT) examination was conducted. Osteonecrosis in the medial sesamoid bone were detected (Figure 1). In subsequent magnetic resonance imaging (MRI), signal changes consistent with osteonecrosis were found (Figure 2).

Upon diagnosis of osteonecrosis, a right short-leg cast was applied. A treatment regimen including naproxen 750mg/day (10 days) and vitamin C 500mg/day (50 days) was started. Contrast bath therapy for the foot and ankle region, transcutaneous electrical nerve stimulation (TENS) (20 min/day), and exercises (range of motion exercises, progressive resistance exercises, balance coordination exercises) were added to the treatment regimen of the patient whose complaints diminished after twenty days of casting. The patient did not have any complaints after 2 months of follow-up.

Case Report 2

A thirty-two-year old female patient admitted to our outpatient clinic for pain in the right sole that had lasted for about one month. Pain significantly increased in the course of each day and was partially relieved with rest. The patient was hardly stepping on the right foot. Three months previously, she had suffered a right ankle anterior talofibular ligament injury due to an inversion sprain when walking, and had received rehabilitation for 3 months. She was evaluated two times by different

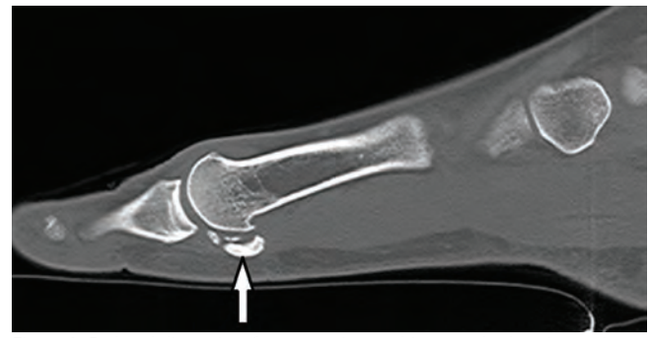

Figure 1. Reduced dimension, fragmentation, and increased sclerosis consistent with osteonecrosis are shown in the medial sesamoid bone in sagittal reformatted computed tomography image.

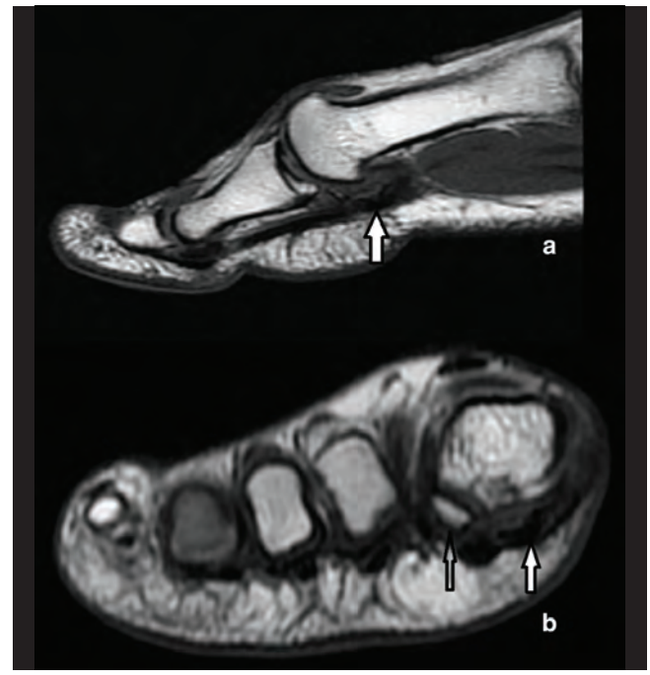

Figure 2. Decreased dimension and signal consistent with osteonecrosis are observed in the medial sesamoid bone (white arrow) in T1-weighted magnetic resonance imaging performed in sagittal (a) and coronal (b) planes. Lateral sesamoid bone (b) is of normal size and normal signal intensity (hollow arrow).

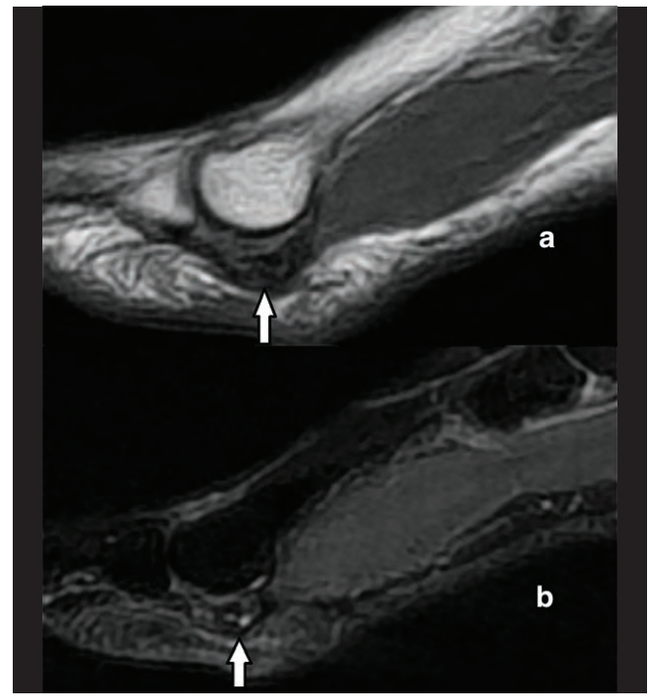

Figure 3. Decreased dimension and signal consistent are observed in the medial sesamoid bone in T1-weighted sequence (a) performed in the sagittal plane in magnetic resonance imaging (white arrow). Occasional hyperintense signals are observed in the osteonecrotic sesamoid bone in STIR sequence (b).

physicians, and took various medications for plantar fasciitis.

In her physical examination, extension of the right MTF joint was significantly limited and painful. The plantar surface of the joint was oversensitive to pressure. There was no increased temperature or swelling. The patient needed to step on the outer surface of the right foot due to pain while walking and had difficulty when walking uphill. Laboratory examinations and direct roentgenogram were normal. In CT examination of the foot performed due to potential fracture, fragmentation and increased sclerosis were detected in the medial sesamoid bone. In the subsequent MRI examination, decreased size and signal were observed in the medial sesamoid bone consistent with osteonecrosis (Figure 3).

The patient was diagnosed with osteonecrosis of the medial sesamoid bone and recommended treatment was meloxicam 15 mg/day, rest, orthopedic insole to prevent weight transfer to the metatarsal region, and contrast bath therapy. The patient's pain had significantly eased by the time of the control examination performed 1 month later and she was admitted to outpatient follow-up.

Discussion

The hallucal sesamoid complex is comprised of medial (tibial) and lateral (fibular) hallux sesamoids. These are functionally important structures. This complex absorbs a great portion of the weight in gait and supports plantar flexion force of the first MTF joint [5, 6].

One of the vascular sources in the circulation of sesamoid bones enters the bone from the proximal end of the insertion of the flexor hallucis brevis tendon and the other enters the bone through the plantar surface of both sesamoids. Another minor source reaches the bone from the distal plantar surface of both sesamoids. Hence, the distal ends of sesamoids have poor circulation support, and the number of these arterial branches affects both bone recovery and avascular necrosis incidence [3, 6]. The lateral sesamoid bone may shift to the first space during the weight-bearing stage and is exposed to less pressure. Thus some publications report that avascular necrosis appears more frequently in the medial sesamoid, while other resources argue that medial and lateral sesamoids are equally affected [3,7]. In both of our patients, there was medial sesamoid involvement.

Foot pathologies associated with sesamoids account for 9% of foot and ankle injuries [7]. Young adults, like the patients described in our case reports, and women are more frequently affected. The contributing factors are generally structural foot defects such as pes cavus and hallux valgus that increase micro trauma risk, and sports activities [8,9]. Repetitive stress, presence of cavus deformity, and exposure to excessive plantar flexion are also listed among the causes of sesamoid injury and avascular necrosis [3]. Post-traumatic sesamoid bone fracture is also a cause of osteonecrosis [10]. It is suggested that, in non-traumatic cases, spontaneous osteonecrosis develops secondary to microvascular events where arterial anastomoses are insufficient or arterial support is deformed [8]. Our second case had a history of trauma, but our first case was consistent with spontaneous non-traumatic osteonecrosis.

The most frequent symptom is pain. Pain occurs with the extension of the hallux in the final phase of the gait cycle when weight is given on the first metatarsal bone. It may have a sudden or insidious onset, and it may become chronic or intermittent. Supination may occur when walking. Pressure on the sesamoid bone, forced plantar, or dorsal flexion of the first toe may result in pain. Inflammatory findings may be noted in the region around the first MTF joint [3,7,10]. In both of our cases, pain was a major complaint as described in the literature.

In imaging studies, roentgenograms should be performed initially [11]. No pathological findings are noted in direct roentgenograms in the acute phase. In serial roentgenograms, radiological changes such as heterogeneous striated sclerosis between 6-12 months, flattening of the sesamoid bone, and demineralization may occur [3,10]. CT is useful as it shows heterogeneous bone structure and abnormal shape and fragmentation. These changes may be noted at the onset of the necrotic process [11]. Scintigraphic studies are useful in diagnosis as increased radionuclide uptake occurs at an earlier phase compared to radiological changes [10]. It should, however, be noted that there may be decreased uptake or no uptake in the early phase [8]. In our patients, findings likely to be consistent with osteonecrosis were detected in CT performed in order to rule out fracture, and definitive diagnosis was made with MRI. The most useful examination method in early diagnosis is MRI because the osteonecrotic process can be monitored [8]. It is also useful in distinguishing other lesions, such as arthritis, chondromalacia, osteochondritis, and stress fracture that might be caused by chronic stress on the hallux sesamoids [12]. Ultrasonography may be helpful for distinguishing pathologies like Morton's neuroma and intermetatarsal bursitis that may cause front leg pain [13]. Diagnostic local anesthetic injections to the MTF joint may be recommended to differentiate intraarticular and extraarticular pathologies. However, this method may prove insufficient in differentiating intrinsic sesamoid pathologies and adjacent soft tissue pathologies like tendinopathies [14].

The basic treatment approach is to reduce the weight on the foot and to use metatarsal or sesamoid pads and arch supports beneath the metatarsal head to relieve the pressure in the regions under pressure. Anti-inflammatory treatments like nonsteroidal anti-inflammatory drugs (NSAIDs), hydrotherapy, cold, ultrasound, and local injections are also useful. In severely symptomatic patients, continuous use of orthosis or a cast may be considered, as we recommended to our first patient [3,10]. Surgical treatment includes partial or total sesamoidectomies. In cases when careful surgical technique is used, deformities after excision of isolated sesamoids are not frequently noted, but when both sesamoids are excised, mechanic advantages of the flexor hallucis brevis muscle may be reduced, and big toe deformities may occur. To reduce complication risk, it is recommended that surgery should be the preferred treatment only when all conservative treatments are unsuccessful; load-bearing mechanism of the first MTF joint should be preserved to the extent possible [3,5].

In conclusion, sesamoid avascular necrosis is a diagnosis that should be considered in patients who apply with persistent foot pain. In differential diagnosis, serial direct roentgenograms, bone scintigraphy, CT, and MRI are useful in addition to patient history and physical examination. Where conservative treatments prove insufficient, surgical treatment can be considered.

Competing interests

The authors declare that they have no competing interests.

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Endometrial Osseous Metaplasia And Infertility: Case Report

Endometriyal Osseöz Metaplazi ve İnfertilite: Olgu Sunumu

Muzeyyen Uyanık[1], Murat Erdemır[2], Ergun Uyanık[3], Safak Atahan[4]
[1]Department of Obs&Gyn, VM Medical Park Hospital Bahçeşehir University, İstanbul [2]Department of Obs&Gyn, Jinemed IVF Center, Bursa,
[3]Department of Obs&Gyn, Yuksek Ihtisas Training and Research Hospital, Bursa, [4]Department of Pathology, Biruni University School of Medicine, Istanbul, Turkey

Özet

Endometriyal osseöz metaplazi infertiliteye sebep olabilen nadir bir durumdur. Sıklıkla geç abortuslardan sonra görülebilmektedir. Etyolojisi net olarak bilinmemekle birlikte teorilerle açıklanmaktadır. Bu olguda; 36 yaşında , nullipar hasta polikliğimize çocuk istemi ile başvurdu. Transvajinal ultrasonografi ile değerlendirmede endometrial alanda rahim içi araç (IUD) benzeri ekojen görünüm saptanması üzerine histeroskopi ile tanı ve tedavisi sağlanmış ,spontan olarak gebe kalmış bir hastayı sunmayı amaçladık. Histeroskopik inceleme ile nadir bir infertilite sebebi olan osseöz metaplazi tanısı konulabilmekte ve histeroskopik yöntem ile kemik parçalarının uzaklaştırılması sonucu fertilite geri dönebilmektedir.

Anahtar Kelimeler

Endometriyal Osseöz Metaplazi; İnfertilite; Histeroskopi

Abstract

Endometrial osseous metaplasia is a rare condition which may cause infertility, and can be seen by following late abortions. Its etiology has not been clearly established, although there are several theories. Herein, we report a 36-year old nullipara patient who was admitted to our clinic with the desire to have a child. She was treated with hysteroscopy based on the presence of an echogenic appearance similar to an intrauterine device (IUD) on the endometrial area as assessed through transvaginal ultrasonography and she subsequently had a spontaneous pregnancy. Osseous metaplasia, which is a rare cause of infertility, can be diagnosed through hysteroscopic examination, and fertility can be restored by removing the bone fragments through hysteroscopy.

Keywords

Endometrial Osseous Metaplasia; Infertility; Hysteroscopy

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Corresponding Author: Muzeyyen Uyanık, Department of Obs&Gyn. VM Medical Park Hospital, Osmangazi, Bursa, Turkey.
GSM: +905438090882 E-Mail: drmuzeyyenuyanik@gmail.com

Introduction

Endometrial osseous metaplasia is a rare condition which may cause infertility. It may present with pelvic pain, dyspareunia, and menstrual irregularities. It may be seen following late abortions. Its etiology is not clearly known; however, endometrial tissue osseous metaplasia or fetal bone retention are considered as the possible causes [1,2]. Herein, we report a case who was admitted to our clinic due to infertility and diagnosed and treated through hysteroscopy.

Case Report

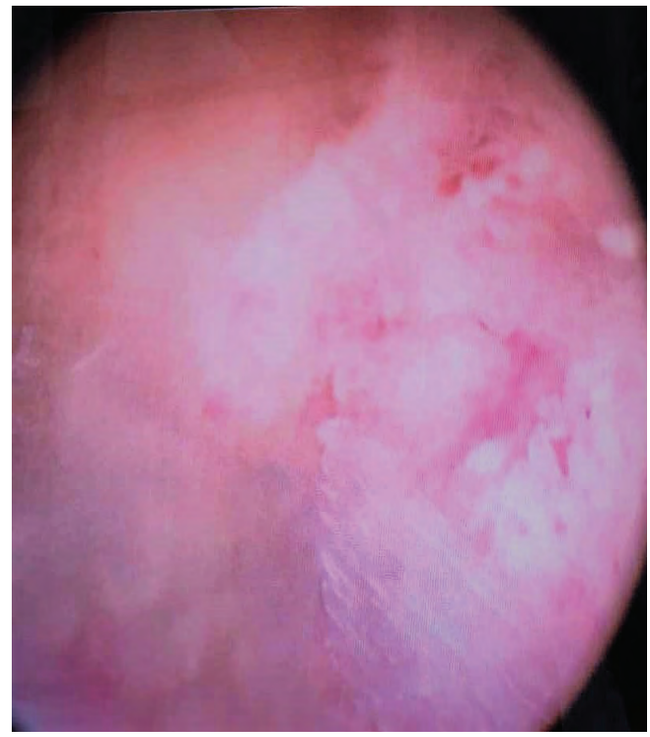
A 36-year old nullipara patient was admitted to our clinic with the desire to have a child. In the third day of menstruation of the patient, transvaginal ultrasonography (TVUSG) revealed normal findings on the endometrial area, except for an echogenic appearance similar to an intrauterine device (IUD) (FSH: 7.2mIU/mL LH: 4.3mIU/mL E2: 58pg/mL spermiogram: normospermia). In the hysteroscopy, excision was performed by following the presence of fine, calcified, and white bone plaques in the endometrial cavity (Picture 1). The entire cavity was, then curetted with a sharp curette. In the histological evaluation, bone trabeculae with a metaplastic mature appearance within the pseudostratified columnar epithelium laid endometrial gland structures was observed, and pathological result was reported as endometrial osseous metaplasia (Picture 2). The patient had a spontaneous pregnancy following hysteroscopic diagnosis and treatment.

Discussion

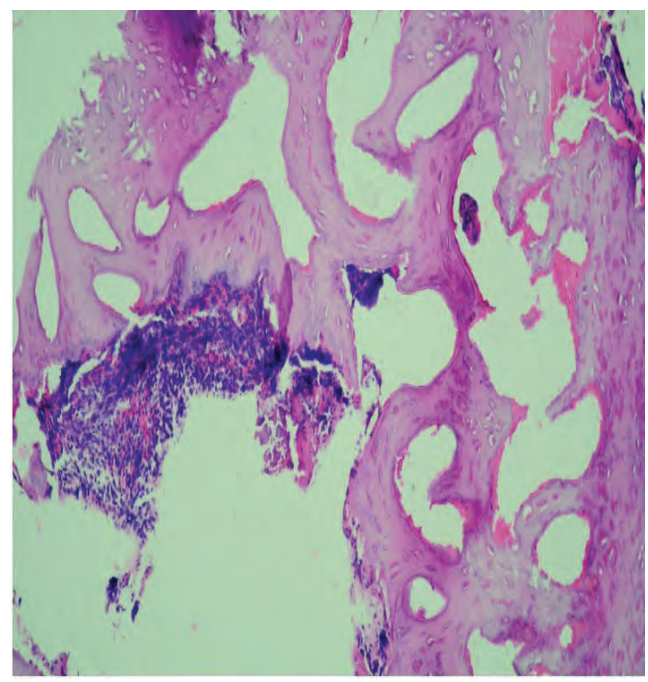
Endometrial osseous metaplasia is a rare condition characterized by abnormal bone formation in the endometrial cavity [1]. Although its etiology still remains unclear, there are two theories regarding the pathophysiology of the disease including: fetal bone retention following an abortion later than three -month gestational weeks, and osseous metaplasia of endometrial stroma [1,2]. Inconsistent with the reported cases in the literature, our case had no abortus history. Possible mechanisms of metaplasia can be defined as the conversion of chronic inflammatory dystrophic calcification to bone formation or myeloid metaplasia, which can be observed in the myometrial transitional zone. Bone fragments in the endometrial cavity are also believed to prevent blastocyst implantation, inducing reactive endometritis as an IUD and leading to infertility [3].

In addition to infertility, endometrial osseous metaplasia may present with several findings such as pelvic pain, dyspareunia and menstrual irregularities [2]. It is often suspected in infertile cases due to the presence of calcification similar to an IUD appearance during the ultrasonographic examination. The diagnosis is made based on the presence of osteoid formations in hysteroscopy and histopathological examination of the specimens; obtained, and fertility can be restored by the removal of bone fragments [4]. Furthermore, Garcia Lean et al. [5] and Rodrigues et al. [6] suggested using laparoscopy as well as hysteroscopic approach to reduce the risk of perforation and to investigate other causes of infertility.

In conclusion, endometrial evaluation in infertile patients is of utmost importance. Hysteroscopy in endometrial evaluation is one of the most useful methods used in daily clinical practice. Osseous metaplasia can be diagnosed and treated through hysteroscopic examination and excision.



Picture 1. Bone Fragments (Hysteroscopy)



Picture 2. Bone trabeculae with a metaplastic mature appearance within the pseudostratified columnar epithelium

Competing interests

The authors declare that they have no competing interests.

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Case Report
JCAM

Prenatal Diagnosis of Congenital Cystic Adenomatoid Malformation of the Lung

Prenatal Tanılı Konjenital Kistik Adenomatoid Malformasyonu

Congenital Hyperechogenic Lung Diseases

Ersen Ertekin[1], Özüm Tunçyürek[1], Figen Tunalı Türkdoğan[1], Mustafa Gök[1], Canten Tataroğlu[2]
[1]Radyoloji AD, [2]Adnan Menderes Üniversitesi, Tıp Fakültesi, Patoloji AD, Aydın, Türkiye

Öz

Konjenital Kistik Adenomatoid Malformasyon, terminal solunum yolu bronşiollerinin hamartomatöz proliferasyonudur ve konjenital akciğer hastalıklarının dörtte birini temsil eder. Prenatal ultrasonografide uniform hiperekoik kitle, değişken ekojenik kistler veya ekojenik stroma ile çevrili multikistik kitle şeklinde görülebilir. 20. hafta ikiz gebeliği olan 30 yaşındaki bayan olgumuzun ultrason incelemesinde, fetal sol hemitoraksı tamamen dolduran ve orta şiddette mediastinal kaymaya neden olan hiperekoik kitle lezyonu gözlendi. Sağ akciğer normal sonografik yapıda idi. Plevral efüzyon veya ek patoloji saptanmadı. Bu olgu sunumu, Konjenital Kistik Adenomatoid Malformasyonun prenatal ultrason bulguları eşliğinde hiperekojen akciğer hastalıkları nedenlerini gözden geçirmiştir.

Anahtar Kelimeler

Konjenital Kistik Adenomatoid Malformasyon, Prenatal Tanı, Hiperekojen Akciğer Hastalıkları

Abstract

Congenital cystic adenomatoid malformation (CCAM) is the hamartomatous proliferation of terminal respiratory bronchioles and represents a quarter of congenital lung diseases. On prenatal ultrasonography, uniform hyperechoic mass, variable echogenic cysts, or multicystic mass surrounded by echogenic stroma may be observed. This case report illustrates prenatal diagnosis of a microcystic-type CCAM in a 30-year-old female patient, pregnant with twins (week 20), and confirmed by the microscopic examination of autopsy. The US scan showed a hyperechoic mass filling the left hemithorax completely and causing a moderate mediastinal shift. The right lung was of a normal sonographic structure, and there was no pleural effusion or additional pathology detected. This report has reviewed the causes of hyperechogenic lung diseases of neonatals with a presentation of the prenatal US findings of a case of CCAM.

Keywords

Congenital Cystic Adenomatoid Malformation, Prenatal Diagnosis, Hyperechoic Lung Diseases

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Corresponding Author: Ersen Ertekin, Radyoloji AD. Adnan Menderes Üniversitesi, Tıp Fakültesi, 09010 Aydın, Türkiye.
GSM: +905332124478 E-Mail: drersen@hotmail.com

Introduction

Congenital cystic adenomatoid malformation (CCAM) is the hamartomatous proliferation of terminal respiratory bronchioles and represents a quarter of congenital lung diseases. Almost 95% of the CCAMs are unilateral. Usually, it is associated with the tracheobronchial tree. As a result of the prevention of alveolar development, cysts occur in the lungs. Cysts are covered by respiratory epithelium [1,2]. Many patients present as respiratory distress in the newborn period. Around 10% of the CCAMs cause recurrent pneumonia with age, while some cases may remain asymptomatic. Many patients present as respiratory distress in the newborn period. Around 10% of the CCAMs cause recurrent pneumonia with age, while some cases may remain asymptomatic. There are three types of CCAMs, which are indicated by the size of the cysts, named macrocystic, microcystic, and mixed-type CCAM [3,4]. On prenatal ultrasonography, uniform hyperechoic mass (microcystic type), variable echogenic cysts (macrocystic type), or multicystic mass surrounded by echogenic stroma (mixed type) may be observed. Cystic masses, especially the microcystic type, can cause mediastinal shift. In this case report, we review prenatal findings of congenital cystic adenomatoid malformation (CCAM) of the lung and hyperechoic lung diseases for which different diagnoses should be considered.

Case Report

A 30-year-old female patient, pregnant with twins (week 20), was admitted to our ultrasound (US) unit for a congenital anomaly scan. The US scan showed a hyperechoic mass filling the left hemithorax completely and causing a moderate mediastinal shift [Figure 1]. The vascularization of the lesion was through the right pulmonary artery. The left lung was of a normal sonographic structure, and there was no pleural effusion or additional pathology detected. According to these findings, we considered a diagnosis of microcystic-type CCAM. Because hydrops and polyhydroamnios were not observed, we recommended close monitoring.

It is not possible to comment on the course of the disease prenatally, as the patient did not come to follow-up examinations. The mother gave birth with caesarian section (C/S) in the 25th gestational week. The male infant with advanced growth retardation was intubated due to cyanosis, and dead on the second day of the follow-up, due to the development of hyaline membrane disease.

In the microscopic examination of autopsy, widely hemorrhagic areas and eosinophilic hyaline membrane formations in the alveoli were observed in both lungs [Figure 2a]. In addition, dilated cystic structures covered by respiratory epithelium similar to bronchus were observed in the lower lung lobe of the left lung [Figures 2b, c, d]

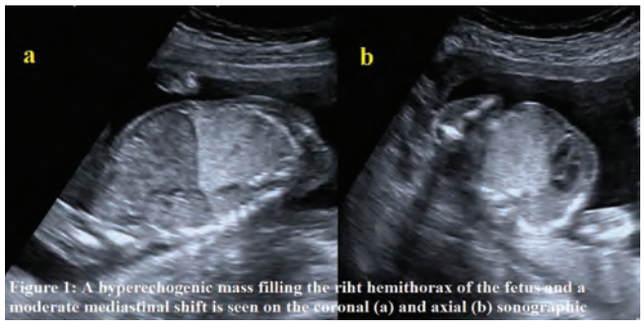

Figure 1. A hyperechogenic mass filling the left hemithorax of the fetus and a moderate mediastinal shift is seen in the coronal (a) and axial (b) sonographic images

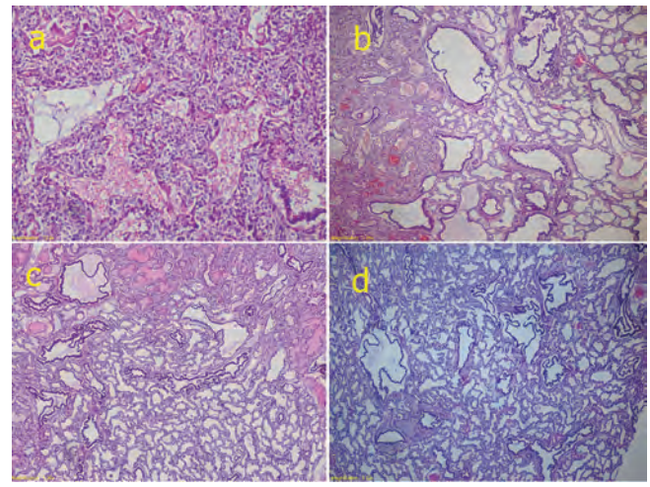

Figure 2. Eosinophilic hyaline membranes in the alveoli (a) and dilated cystic structures covered by respiratory epithelium (b, c, d) are seen in pathological specimens

Discussion

CCAM is the hamartomatous proliferation of terminal respiratory bronchioles and represents a quarter of congenital lung diseases. Almost 95% of the CCAMs are unilateral. Usually, it is associated with the tracheobronchial tree, but vasculatory structures are normal. As a result of the prevention of alveolar development, cysts occur in the lungs. Cysts are covered by respiratory epithelium [1,2]. As in our case, many patients present as respiratory distress in the newborn period. Around 10% of the CCAMs cause recurrent pneumonia with age, while some cases may remain asymptomatic.

There are three types of CCAMs, which are indicated by the size of the cysts, named macrocystic, microcystic, and mixed-type CCAM [3,4]. The best prognosis is where one or more cysts larger than 1 cm are observed (macrocystic CCAM), while solid-looking microcysts indicate the worst prognosis type (microcystic CCAM). On prenatal ultrasonography, uniform hyperechoic mass (microcystic type), variable echogenic cysts (macrocystic type), or multicystic mass surrounded by echogenic stroma (mixed type) may be observed. Cystic masses, especially the microcystic type, can cause mediastinal shift. While macrocystic and mixed types of CCAM may persist during pregnancy, regression is observed in the majority of microcystic CCAM cases [3,5].

Hydrops and polyhydroamnios are common phenomena in microcystic CCAM, as compared to the other types of CCAM. In-utero or postpartum exitus is observed in the vast majority of hydrops cases [6].

For differential diagnosis, the other reasons for hyperechogenic lung (congenital high airway obstruction syndrome [CHAOS] and pulmonary sequestration [PS]) should be kept in mind [1,3]. Display of the feeding arteries from the aorta is diagnostic for PS, although PS cannot be excluded just because it is not shown [2]. In CHAOS, hyperechogenic masses are seen in the bilateral hemithorax, while the mediastinum and hearth are compressed due to the massive enlargement of the lungs [7].

The treatment for microcystic CCAM is open lobectomy, although US guided laser ablation of the feeding arteries can be applied. In macrocystic and mixed-type CCAMs, a thoracoamniotic shunt may be performed, which has a success rate of 2/3 [4,8].

In our case, in accordance with the literature, a uniform hyperechogenic mass filling the left hemithorax and displacing the mediastinum to the right was observed on US examination, and respiratory distress appeared after the birth. The premature birth and low birth weight of the infant led to the development of hyaline membrane disease and, as a consequence of that, the clinical condition declined further.

This report has reviewed the causes of hyperechogenic lung diseases of neonatals with a presentation of the prenatal US findings of a case of CCAM.

Competing interests

The authors declare that they have no competing interests.

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Case Report
JCAM

Effectiveness of Exchange Transfusion in Hyperlipoproteinemia Type 1

Hiperlipoproteinemi Tip 1'de Exchange Transfüzyonun Etkinliği

Pembe Soylu Ustkoyuncu[1], Mustafa Kendirci[1], Songül Gökay[1], Fatih Kardas[1], Ismail Dursun[2], Tamer Günes[3]
[1]Department of Pediatrics, Division of Pediatric Nutrition and Metabolism, [2]Department of Pediatrics, Division of Pediatric Nephrology, [3]Department of Pediatrics, Division of Neonatology, Erciyes University, School of Medicine, Kayseri, Turkey

Öz

Hiperlipoproteinemi Tip–1, lipoprotein lipaz (LPL) eksikliği sonucunda ortaya çıkan, lipoprotein metabolizmasının bir bozukluğudur. Akut pankreatite bağlı tekrarlayıcı karın ağrısı, eruptif ksantomlar, hepatosplenomegali ve lipemia retinalis hastalığın önemli klinik bulguları arasındadır. 40 günlük erkek hasta ishal, beslenememe, kusma ve sürekli ağlama şikâyetleri ile başvurdu. Plazmasının beyaz kremsi görünümde olması nedeniyle lipid düzeyleri çalışıldı. Plazma trigliserit (TG) düzeyi 47450 mg/dl, total kolesterol düzeyi ise 1614 mg/dl idi. Teknik problemler nedeniyle plazmaferez uygulanamadı. Alternatif tedavi olarak exchange transfüzyon (ET) uygulandı. TG düzeyi ET'den sonra ilk saatte 47450 mg/dl'den 3791 mg/dl'ye düşürüldü ve yağdan fakir diyet ve omega-3 yağ asidi desteği ile 300 mg/dl civarında tutuldu. Plazmaferez teknik olarak imkânsız olduğunda, plazmaferezin alternatif tedavisi olarak şiddetli hipertrigliseridemisi olan infantlara exchange transfüzyon uygulanabilir.

Anahtar Kelimeler

Exchange Transfüzyon; Hiperlipoproteinemi Tip–1; LPL Geni

Abstract

Hyperlipoproteinemia Type-1 is a disorder of lipoprotein metabolism caused by the deficiency of lipoprotein lipase (LPL). Recurrent abdominal pain due to acute pancreatitis, eruptive xanthomas, hepatosplenomegaly, and lipemia retinalis are major clinical findings of the disorder. A 40-day-old boy presented with diarrhea, poor feeding, vomiting, and constant crying. Due to the white creamy appearance of his plasma, lipid levels were obtained. His plasma triglyceride (TG) level was 47450 mg/dl and total cholesterol level was 1614 mg/dl. Plasmapheresis was not applied because of the technical problems. Exchange transfusion (ET) was performed as an alternative therapy. The TG level was reduced from 47450 mg/dl to 3791 mg/dl in the first hour after ET and it was maintained at around 300 mg/dl by treatment with a low-fat diet and supplementation of omega-3 fatty acid. Exchange transfusion can be applied to infants with severe hypertriglyceridemia as an alternative treatment to plasmapheresis when plasmapheresis is technically impossible.

Keywords

Exchange Transfusion; Hyperlipoproteinemia Type-1; LPL Gene

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Corresponding Author: Pembe Soylu Ustkoyuncu, Department of Pediatrics, Division of Pediatric Nutrition and Metabolism, Erciyes University, School of Medicine, 38039, Melikgazi, Kayseri, Turkey. T.: +90 3522076666/25280, F.: +90 3524375825 E-Mail: drpembesoylu@erciyes.edu.tr

Introduction

Lipoprotein lipase (LPL) deficiency, also known as familial chylomicronemia or Hyperlipoproteinemia Type-1, causes lipolysis deficiency and chylomicron accumulation. It is a rare, autosomal recessive inherited inborn error of metabolism that occurs in early infancy or childhood. Fasting serum triglyceride (TG) levels are usually above 1000 mg/dl [1].

The disorder occurs due to homozygous or compound heterozygous mutations in five different genes. These genes are *LPL, APO-C2, APO-A5, GPIHBP1,* and *LMF1*. LPL deficiency occurs in 95% of LPL mutations with a frequency of 1/1000000 [1,2].

Growth retardation, eruptive xanthomas, lipemia retinalis, hepatosplenomegaly, and recurrent abdominal pain due to acute pancreatitis are major findings of the disorder. Plasmapheresis is the most commonly used treatment option in LPL deficiency [3].

Here we present a case of LPL deficiency with severe TG elevation in which exchange transfusion (ET) was used successfully, without any complication.

Case Report

A 40-day-old boy presented with complaints of poor feeding, constant crying, diarrhea, and vomiting. In physical examination, his body weight was 4300g (25-50% percentile), height was 54cm (25-50% percentile), and head circumference was 38cm (25% percentile). He was born weighing 3500 grams by ceserean section, at term, the first child of a non-consanguineous Turkish couple.

He was referred to our hospital due to the white creamy appearance of his plasma and his lipid levels were obtained. Plasma TG level was 47450 mg/dl (N: 35-150), total cholesterol level was 1614 mg/dl (N: 70-200), and HDL cholesterol level was 89 mg/dl (N: 0-90). LDL cholesterol level was not measured. Liver-renal function tests, amylase, lipase levels, and abdominal ultrasonography were normal.

Plasmapheresis was not applied because our hospital's apheresis device is not suitable for use in children under 25 kg. For this reason, we decided to perform exchange transfusion (ET) by attaching a central venous catheter. After informed consent was obtained from the family, ET was performed using 180 ml of whole blood per kg.

Blood glucose, electrolytes, complete blood count, and liver and renal function tests were within normal limits after the ET. Heart and respiratory rates and arterial blood pressure were within normal ranges. Within the first hour after ET, the TG level was reduced from 47450 mg/dl to 3791 mg/dl. The TG and total cholesterol levels of the patient following ET are shown in Table 1.

48 hours after the procedure, oral feeding was started with a formulation containing 8% fat (50% consisted of medium chain triglycerides), 15% protein, and 77% carbohydrate.

Table 1. The TG and total cholesterol levels of the patient after ET

Time	TG level (mg/dl)	Total cholesterol level (mg/dl)
First hour after ET	3791	381
12th hour after ET	1710	310
24th hour after ET	1178	254

Abbreviations: TG: Triglyceride, ET: Exchange Transfusion

On day 15, TG levels were measured as 629 mg/dl, total cholesterol was 251 mg/dl, HDL cholesterol was 17 mg/dl, and LDL cholesterol was 107 mg/dl. 2 g/day of omega-3 fatty acid was added to his diet.

The TG level was maintained around 300 mg/dl by treatment with a low-fat diet and supplementation of omega-3 fatty acid. Molecular genetic analysis of the patient was performed. A compound heterozygous mutation [p. T211A (c.613A>G)/p. R270C (c.808C> T] was identified in the *LPL* gene and lipemia retinalis was detected in his ophthalmological examination.

Discussion

Seizures, encephalopathy, pancreatitis, severe psychomotor retardation, spasticity, and blindness are some of the complications associated with severe TG elevations. Wilson et al. [4] reported a 5-week-old infant with encephalopathy due to lipid deposition in the brain. Spasticity and severe psychomotor retardation were observed at 6 months old, and blindness at 18 months in this patient.

The use of a low-fat diet and omega-3 fatty acids causes a decrease in TG levels in the treatment of patients with hypertriglyceridemia, but it may not be sufficient alone to achieve a rapid reduction of severe TG elevations. Therefore, emergency treatment should be done to avert a severe pancreatitis episode and/or other complications.

Plasmapheresis is one of the treatment option for cases with severe TG elevation. Different plasmapheresis techniques have been used in treatment of adult patients with severe TG elevation since 1978. The beneficial effect of plasmapheresis is believed to be due to the removal of excessive proteases from the plasma and replacement of consumed protease-inhibitors with new ones from donor plasma. Plasmapheresis is limited in infants due to hemodynamic effects and hemorrhagic events caused by extracorporeal procedures. The plasma filtration technique is preferred by some centers because it requires less extracorporeal circulating fluid volume. The indications for the use of plasmapheresis are medical emergencies such as pancreatitis with excessively elevated TG levels (TG>1000 mg/dl) [3]. Although our patient did not have pancreatitis, he had poor feeding, vomiting with diarrhea, and was constantly crying. Stefunutti et al. [5] reported a 3-month-old patient with severe TG elevation who had plasmapheresis without pancreatitis, similar to our case.

Exchange transfusion has been used extensively since the 1940s to prevent the development of kernicterus at severe indirect bilirubin elevations [6]. In the literature, very few patients have been shown to have had an exchange transfusion due to severe triglyceride elevation.

Önal et al. [7] reported a 6-month-old patient with resistant myoclonic and tonic seizures who had a lipoprotein lipase deficiency. The seizures were not controlled with infusion of phenobarbital and midazolam. This patient's TG level was 51300 mg/dl. It was not decreased with medical treatments and so, due to the technical problems in plasmapheresis, exchange transfusion was performed. In our case we chose exchange transfusion for a similar reason and we observed a significant decrease in triglyceride levels.

Various complications can be seen after an exchange transfusion. The most common complications are thrombocytopenia, hypocalcemia, hyperkalemia, apnea, bradycardia, hypotension, encephalopathy, and catheter-related events. Steiner et al. [6] reported that transfusion-related complications were transient. Complications were observed most often in preterm and/or very ill newborns. None of these complications developed after the exchange transfusion in our case.

In conclusion, it is necessary to determine treatment methods that can be applied in the acute period before medical treatment in cases with severe TG elevation. We consider that most of the complications related to exchange transfusion are temporary. Due to the hemodynamic effects and hemorrhagic events of plasmapheresis, and for low-weight children who cannot receive plasmapheresis, exchange transfusion may be performed.

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Author Contributions

PSU and MK designed the case report. PSU prepared the manuscript. SG and FK analyzed and evaluated the data. ID and TG provided health care to the patient. The final manuscript was approved by all of the authors.

Conflict of interest

The authors have no financial or personal relationships that could pose any conflict of interest.

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Ethical approval

Our single case report does not require ethics committee approval. Written consent was obtained from the parents.

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The Case of Frontal Linear Scleroderma (En Coup De Sabre)

Frontal Lineer Skleroderma (En Coup De Sabre) Olgusu

En Coup De Sabre

Gülhan Gürel[1], Sevinç Şahin[2], Emine Çölgeçen[1]
[1]Department of Dermatology, [2]Department of Pathology, Bozok University School of Medicine, Yozgat, Turkey

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Öz

En coup de sabre terimi frontal ya da frontoparyetal bölgede özel bir görünüme sahip olan linear skleroderma'nın nadir bir varyantıdır. Etiyolojisi net olarak bilinmemekle birlikte otoimmün kaynaklı olabilir. Nörolojik ve oftalmolojik bulgularla beraber olabilen bu hastalık çocuklarda daha sık görülmektedir. On sekiz yaşında kadın hasta polikliniğimize yaklaşık 4 yıldır alın orta hatta çöküntü ve renk değişikliği şikayetiyle başvurdu. Dermatolojik muayenesinde frontal bölgede orta hattan saçlı deriye doğru uzanan atrofik, lineer , deprese plak lezyonu mevcuttu. Hastaya mevcut klinik ve histopatolojik bulgularla ' linear skleroderma (En coup de sabre)' tanısı konuldu. Hastanın yapılan göz muayenesinde patolojik bulguya rastlanmadı. Nörolojik muayenesi normal olan hastanın kranial magnetik rezonans incelemesinde beyinde patolojik bulguya rastlanmadı.

Anahtar Kelimeler

En Coup De Sabre; Skleroderma

Abstract

En coup de sabre is a rare variation of linear scleroderma, which has a special appearance in the frontal or frontoparietal region. Although the etiology is not clearly known, it can be an autoimmune origin. This disease, which may be associated with neurological and ophthalmological findings, is more common in children. An 18-year-old female patient was admitted to our polyclinic with complaints of collapse and discoloration on the forehead line for approximately four years. The dermatologic examination revealed an atrophic, linear, and depressed lesion on the frontal region extending to the scalp. The patient was diagnosed with 'linear scleroderma (En coup de sabre)' with clinical and histopathological findings. No pathological findings were found on the ophthalmic examination of the patient. Neurological examination was normal and cranial magnetic resonance imaging revealed no pathological findings in the brain.

Keywords

En Coup De Sabre; Scleroderma

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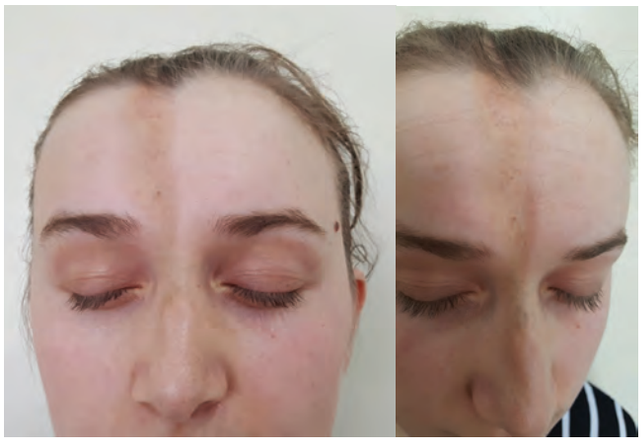
Corresponding Author: Gülhan Gürel, Department of Dermatology, Medical Faculty, Bozok University, 66200 Yozgat, Turkey.
Tel: +90 3542127060 F.: +90 3542177150 E-Mail: gulhanozturkgurel@hotmail.com

Introduction

Morphea (localized scleroderma) is an inflammatory skin disease that causes sclerosis in the dermis and subcutaneous adipose tissue [1]. Linear scleroderma is the most common form of localized scleroderma in children and adults. The term en coup de sabre (ECDS) is used to describe a variant form of linear scleroderma, which has a special appearance in the frontal or frontoparietal region. ECDS emerges in the form of linear, erythematous bands, and gradually transforms into firm plaques. It is usually seen on the paramedian region on the forehead and is unilateral [2]. Here, we present a case of an 18-year-old female patient, who is diagnosed with ECDS after clinical and histological examinations.

Case Report

An 18-year old female patient was admitted to our clinic with collapse and discoloration on the forehead line. The patient's medical history showed that pink-purple color change started approximately four years ago, which was followed by a progressive collapse on the forehead line. The patient's family history did not show anyone with a similar disease. Systemic examination was normal, and physical examination did not show any pathological findings. Dermatologic examination showed that the patient had an atrophic, linear, and depressed lesion on the frontal region extending to the scalp [Figures 1 and 2]. The patient's routine laboratory tests results were within expected intervals. The patient underwent skin biopsy from the frontal region. A written informed consent form was obtained before the procedure.



Figures 1 and 2. Atrophic, linear, and depressed plaque lesion on the frontal region, extending from midline to the scalp.

Histopathological examination showed superficial orthokeratosis and irregular acanthosis in the epidermis. An increase in the number of thick, homogenized, collagen fibers, involving the whole dermis and affecting the subcutaneous adipose tissue, and intermittent atrophy in skin adnexa were present [Figure 3]. Verhoeff-Van Gieson staining (VVG) showed thick, fragmented, elastic fibers laying parallel to the dermis surface [Figure 4].

In light of clinical and histopathological findings, the patient was diagnosed with linear scleroderma (ECDS). The ophthalmic examination did not reveal any pathological findings. The patient's neurological examination was normal, and cranial magnetic resonance imaging (MRI) did not show any pathological findings in the brain.

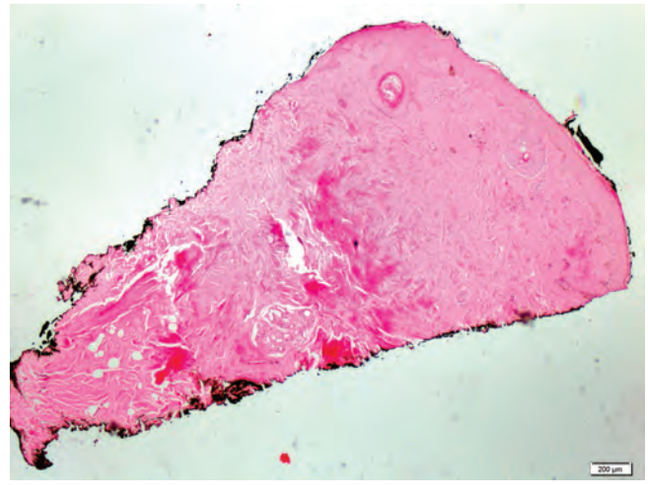


Figure 3. Punch skin biopsy showing loss of skin adnexa, and increased number of homogeneous, collagen fibers in the dermis and subcutaneous adipose tissue (H&E staining, x40).

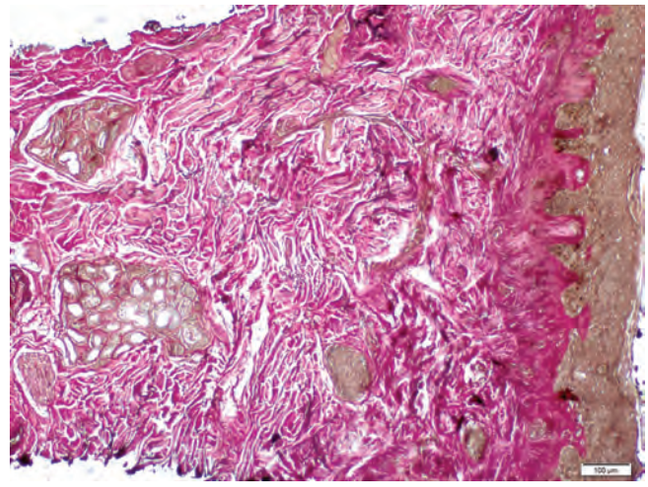


Figure 4. Thick, fragmented, elastic fibers laying parallel to the dermis surface (VVG staining, x100).

Discussion

ECDS is a rare variant of morphea. The disease is usually seen during childhood, and the average age at onset is ten years [2]. A retrospective study on 82 patients indicated that ECDS has an incidence of 0.13/100,000 [3].

Similar to other types of scleroderma, the exact etiology of ECDS remains unknown [2,4]. The disease is considered to have an autoimmune origin and triggered by environmental factors. The exact relationship between progressive facial hemiatrophy (Parry Romberg Syndrome), which is characterized by progressive atrophy on one half of the face, and ECDS is not known. Still, given the similarities in the pathogenesis of these diseases, they are considered to lie on different ends of the spectrum of the same disease [5]. In the present study, our patient did not have facial atrophy.

Neurological involvement with ECDS has been identified in 18-47% of the cases. Epileptic seizures are the most common symptoms, while hemiparesis, muscle weakness, headache, personality changes, and deterioration in intellectual functions have also been reported [6,7]. Our patient's neurological examination was normal, and cranial MRI did not reveal any pathological findings. It is also possible to detect ophthalmologic anomalies in ECDS; however, our patient did not have any

abnormal findings in the ophthalmic examination.

Methotrexate and systemic glucocorticoids represent the first line treatment for ECDS, and it is recommended to maintain treatment with UVA1, PUVA photochemotherapy, narrowband UVB or mycophenolate mofetil. Other treatment options include topical tacrolimus, topical vitamin D and glucocorticoid combinations, and imiquimod [8].

In conclusion, we wanted to present this case as ECDS represents a rare variant of morphea.

Ethical Responsibilities: All institutional and national guidelines for the care and use of laboratory animals were followed.

Conflict of Interest: No potential conflict of interest relevant to this article was reported.

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Competing interests

The authors declare that they have no competing interests.

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Case Report
JCAM

RETINAL HAEMORRHAGE IN A PRETERM NEWBORN – A CLINICAL CASE

Kiril Slaveykov[1], Kalina Trifonova[2], Dimitar Dzhelebov[2], Hristo Mumdzhie[3]
[1]Department of General Medicine, [2]Department of Ophthalmology, [3]Department of Neonatology, Trakia University, Bulgaria

Öz

Amaç: Retinal kanamalar, sağlıklı yenidoğanlarda doğumdan sonraki saatlerde izlenen, sık görülen bir durumdur. Bunların büyük bir yüzdesi, doğumdan sonraki ilk iki hafta içinde rezorbe olmaktadır ve sadece tek bir vakada dört haftadan sonra retinal kanamalar bildirilmektedir. Olgu Sunumu: Hastamız, gebeliğin 26. haftasında doğmuş, 690 gram ağırlığında prematüre bir bebektir. Prematürite Retinofatisi taraması sırasında alakalı herhangi bir bulguya rastlanmayıp, kanama bulundu. Tarama indirekt oftalmoskopi ile yapılmış olup, fotoğraflar ise, iphone'a uyarlı welch-allyn oftaloskop ile çekildi. Kanama 20 haftanın sonunda ancak tamamiyle rezorbe edilebildi. Vakanın retcam ile dökümantasyonu, oftalmoloji bölümüne sevkinden sonra mümkün olmuştur. Tartışma: Kanamaların prematüre ve sezeryanla doğmuş bebeklerde nadiren görülmesine rağmen, çok düşük kiloyla doğan ve yapay havalandırmaya tabi tutulan bebekler için hala büyük bir risk teşekkül etmektedir.

Anahtar Kelimeler

Oftalmoloji; Kanama; Prematüre

Abstract

Aim: Retinal hemorrhages are frequently observed sights in healthy newborns examined hours after birth. A large percentage of them are resorbed in the first two weeks following delivery, with a single case reporting retinal hemorrhages after four weeks. Case Report: We present a preterm newborn weighing 690 g at birth, born at 26 g.w. A hemorrhage was found during screening for retinopathy of prematurity with no signs of retinopathy of prematurity. The screening was performed with indirect ophthalmoscopy, and a photo was taken with a welch-allyn ophthalmoscope adapted to an iphone. The hemorrhage had persisted for nearly 20 weeks before it was completely resorbed. Case documentation with retcam was possible after transportation to the Department of Ophthalmology. Discussion: Even though hemorrhages are less common in children born preterm and with C-section, infants with extremely low birth weight and those who underwent artificial ventilation are still at increased risk for retinal hemorrhages.

Keywords

Ophthalmology; Hemorrhage; Preterm

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Corresponding Author: Kiril Slaveykov, Bulgaria, Stara Zagora, Armeiska 11 str.,
T.: +35988671278 E-Mail: kirilslaveykov@gmail.com

Introduction

Retinal hemorrhages (RH) are frequently observed sights in healthy newborns examined hours after birth. They are more common in full-term babies compared to preterms. A large percentage of them are resorbed in the first two weeks following delivery, with only single cases reporting RH after four weeks. RH in infants older than one month should increase suspicion that the hemorrhage is associated with factors other than birth. The incidence of RH is higher for vacuum-assisted births than spontaneous vaginal deliveries and forceps delivery and is least for infants delivered by cesarean section. RH are observed more often in term compared to preterm neonates, which is due to the greater head circumference leading to fetal head compression and venous congestion during delivery [1].

A study from 2011 focuses on neonates with a medical history of perinatal distress which includes the following: birth asphyxia, meconium aspiration, amniotic fluid aspiration, fetal distress, transient tachypnea of the newborn, and dysphagic choking. Under these conditions, intraocular hemorrhages might result from significant hypoxia [2]. Autoregulatory hypoxic cerebral vasodilatation produces an increase in intracranial pressure, which in turn increases the retinal venous pressure [3]. Aspirated meconium, amniotic fluid, blood, or any source of airway irritation can cause mechanical obstruction. The forceful effort to extrude the irritant material may increase intrathoracic pressure and subsequently lead to cephalic venous congestion [2].

An observational study was carried out on 11 premature infants in whom retinal and/or vitreous hemorrhages had been observed within their first months of life. Contrary to the quick absorption (<1-2 weeks only) usually seen in most newborn term infants, the ocular bleeding in preterms was generally longstanding. The authors suggest that prematurity as such is added to the list of possible underlying causes when retinal bleedings are evaluated in very small infants [4].

Some maternal factors are also discussed by different authors. Maternal smoking during pregnancy causes increased frequency of retinal arterial narrowing and straightening, retinal venous dilatation, and tortuosity and intraretinal hemorrhages [5]. RH occurred with greater frequency in neonates born to women who had an intrauterine infection [1].

Case Report

We present a case of a preterm newborn from a first pathological pregnancy with a p.t.b 01.10.2016. The infant was born with a C-section on 01.07.2016, with 26 g.w. morphological maturity, weighing 690 at birth and severe asphyxia. The infant was intubated on the third minute and extubated on the third day. Subsystemic pulmonary hypertension and neonatal hepatitis were found. At day 11 a clinical picture of bronchopulmonary dysplasia was present, after which bilateral respiratory associated pneumonia developed with frequent episodes of apnea and respiratory failure. After improvement of the condition, treatment with oxygen via tent was given. Artificial ventilation was administered for a total of 15 days, CPAP – 3 days, surfactant – 2 times with oxygen therapy. Due to the severity of the child's condition screening examination for retinopathy of prematurity was possible only in 33 g.w. A hemorrhage was found on the path of the superior temporal arcade of the retinal artery in the left eye during indirect ophthalmoscopy. During follow-up three weeks later (36 g.w.) the hemorrhage blot persisted and was documented as 1.5 – 2 disk diameters large and three disk diameters away from the papilla (Fig 1-2). The screening was performed with indirect ophthalmoscopy, after which a photo was taken with a Welch Allyn iExaminer system (a panoptic ophthalmoscope adapted to an iPhone). During the control exams 38 and 40 g.w. the hemorrhage was still present. In the follow-up at 2 and 4 weeks after term, the hemorrhage started to decrease in size and documentation with a retcam was possible after transportation to the Department of Ophthalmology. Retcam examination was performed six weeks after term, and the hemorrhage was completely resorbed (Fig 3-4). Ophthalmological examination results: pupils equal, round, reactive to light with good mydriasis after Mydfrin 2,5% and Tropicamide 0,5%, lens and vitreous – transparent, papilla – vital, with clear borders, on the retinal level, vessels – with slightly increased tortuosity and normal caliber, reaching 2nd zone. No sign of retinopathy of prematurity was found during the examination.

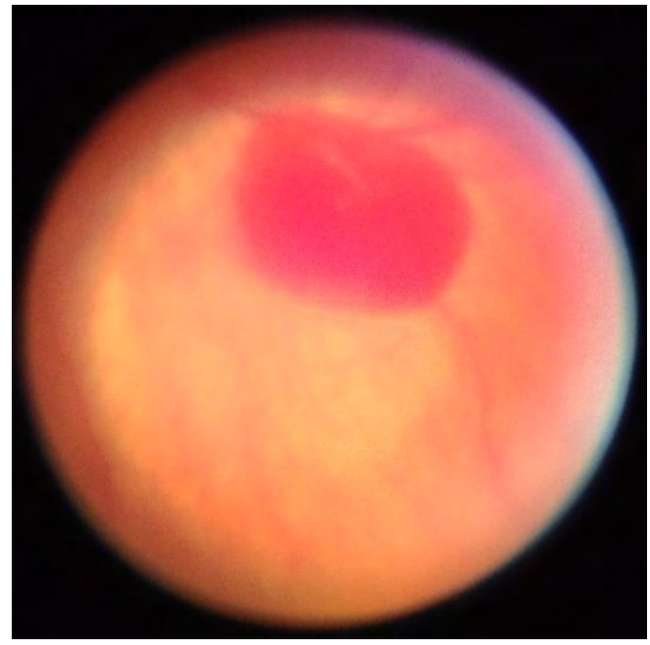

Fig 1. Haemmorhage on the superior temporal arcade of the left eye

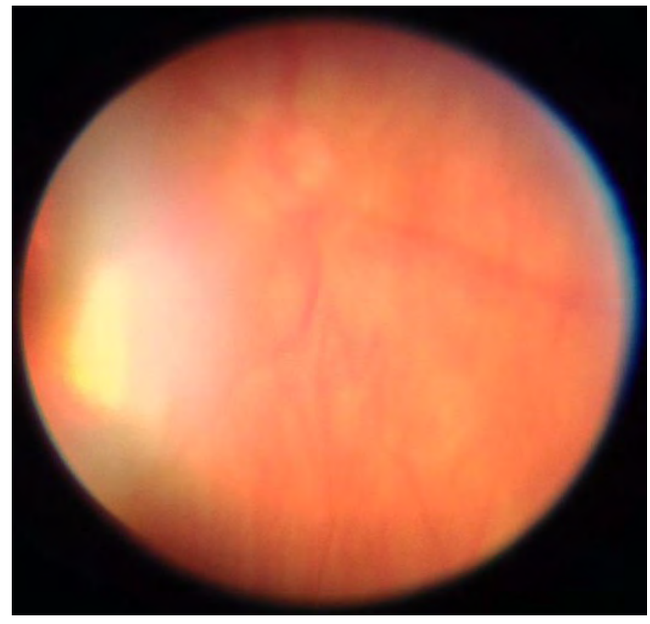

Fig2. Papilla of the left eye

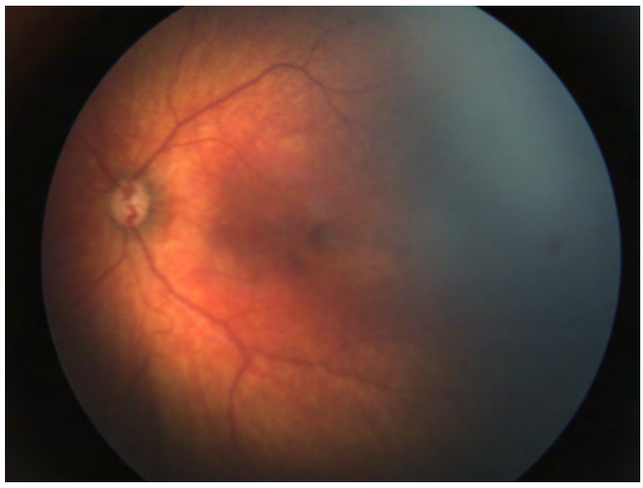

Fig 3. Left eye fundus 6 weeks after term

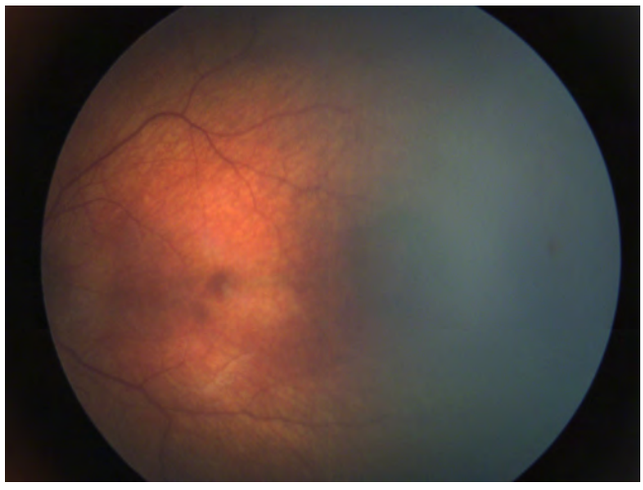

Fig 4. Left eye fundus 6 weeks after term

Discussion

The anatomical location and appearance of RH in the infant provide important clues in the diagnosis of underlying disorders. While neonatal RH related to birth trauma are common, benign, and self-limited, other RH in infancy may signify intracranial aneurysms, accidental or non-accidental injury, and a variety of ocular (e.g., Coats' disease, PHPV, retinopathy of prematurity, retinal dysplasia, hypertension, myopia) or systemic disease (e.g., hematologic or cardiovascular disorders, infection, protein C deficiency) [6].

The broad spectrum of available etiology options presents a challenge and makes differential diagnosis difficult for the screening ophthalmologists.

The most common ocular manifestation of shaking injury, present in a large majority of cases, is RH. They are thought to result from repetitive abrupt deceleration of the child's head as it whiplashes back and forth during the shaking episode. A striking feature of shaken baby syndrome is the typical lack of external evidence of trauma. The ocular adnexa and anterior segment appear entirely normal. Occasionally, the trunk or extremities show bruises representing the imprint of the perpetrator's hands.

Patients with subarachnoid hemorrhage, or other causes of rapidly increased intracranial pressure, may develop ocular hemorrhage (Terson syndrome). Clinical ophthalmologic findings may have significant diagnostic and prognostic value for clinicians [7]. A rapid increase in intracranial pressure secondary to subarachnoid hemorrhage following rupture of an aneurysm can result in sequelae similar to those found in inflicted traumatic brain injury. The case of a 1-month-old girl with bilateral RH resulting from a ruptured cerebral aneurysm is described by Joseph Scheller and Pavle Doroslovacki [8].

RH have been reported as the first manifestation of leukemia. They involve the posterior fundus and can have some correlation with other aspects of the disease such as anemia, thrombocytopenia, or coagulation abnormalities. The RH in leukemia can resemble those associated with intracranial hemorrhages and trauma.

RH can be seen after cardiopulmonary resuscitation but are rarely extensive by the hands of experienced personnel.

Even though we can exclude the most common causes for RH (the child is born pre-term, via C-section, there is no indication of trauma or shaken baby syndrome, leukemia, cerebral aneurysm or increased intracranial pressure), extremely low birth weight remains a primary suspect and is a sufficient factor on its own as a cause of the hemorrhage. Possible secondary factors include artificial ventilation, neonatal hepatitis, neonatal distress, and trauma during surgery. The severe prematurity is also the reason for the prolonged retention of the hemorrhage.

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All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Competing interests:

The authors declare that they have no competing interests.

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Pseudoxanthoma Elasticum: Report of Two Cases

Psödoksantoma Elastikum: İki Olgu Sunumu

Common Eye and Skin Findings in the Pseudoxanthoma Elasticum

Suzan Demir Pektas[1], Omur Demirtas[2], Gulen Gul[3], Yelda Dere[4], Suphi Bulent Sarı[5], Gursoy Dogan[1], Aylin Karalezli[5]
[1]Department of Dermatology, Muğla Sıtkı Kocman University Faculty of Medicine, Mugla,
[2]Department of Ophthalmology, Aydin State Hospital, Aydın, [3]Department of Pathology, Tepecik Training and Researh Hospital, İzmir,
[4]Department of Pathology, Muğla Sıtkı Kocman University Faculty of Medicine, Mugla,
[5]Department of Ophthalmology, Muğla Sıtkı Kocman University Faculty of Medicine, Mugla, Turkey

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Öz

Psödoksantoma elastikum (PKE), elastik bağ dokusunun patolojik kalsifikasyonu ile karakterize genetik geçişli, multisistemik bir hastalıktır. PXE, olguların %90'ında otozomal dominant kalıtım paternine sahip nadir görülen bir hastalıktır. Erkeklere göre kadınlarda daha sık görülür. PKE gelişiminde transmembran transporter proteinini kodlayan ABCC6 (ATP bağlayıcı kaset alt ailesi C elemanı 6) genindeki mutasyonun rol oynadığı gösterilmiştir. Klinik bulgular; genellikle deri, göz, mukoza, gastrointestinal sistem ve arterlerde ortaya çıkar. PXE'de asemptomatik cilt lezyonları, genellikle ilk klinik bulgudur. PXE'de peau d'orange, kuyruklu yıldız lezyonları, anjioid çizgileri, koroid neovaskülarizasyonu (KNV), kanamalar oftalmolojik tutulumla ilişkili başlıca bulgulardır. PXE'li hastalarda nadiren erken erken ateroskleroz, akut miyokard infarktüsü ve serebrovasküler olaylar gelişebilir. Bu makalede, PKE tanılı iki kadın olguyu sunarak hastalığın klinik özelliklerini, hastalıkla ilişkili komplikasyonları ve hastalığın etkin tedavisi için erken teşhisin önemini vurgulamayı amaçladık.

Anahtar Kelimeler

Deri; Göz; Psödoksantoma Elastikum

Abstract

Pseudoxanthoma elasticum (PXE) is an inherited multisystem disorder characterized by pathological calcification of the elastic connective tissue. PXE is a rare disease with an autosomal dominant inheritance pattern in 90% of the cases. Females are more commonly affected than males [2]. PXE is caused by mutations in the ABCC6 (ATP-binding cassette sub-family C member 6) gene that encodes a transmembrane ATP binding efflux transporter. Clinical manifestation occurs in skin, eyes, mucosa, gastrointestinal tract and the arteries. Asymptomatic skin manifestations, which are often the first clinical signs of PXE. Ophthalmological features of PXE include primarily peau d'orange, comet lesions, angioid streaks, choroidal neovascularization (CNV), hemorrhages. The patients of PXE can also develop premature atherosclerosis with early acute myocardial infarcts and cerebrovascular accidents. We reported two female cases of PXE, emphasizing its main clinical aspects and highlighting the importance of early diagnosis of the disease for an adequate therapeutical management of associated complications.

Keywords

Eyes; Pseudoxanthoma Elasticum; Skin

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Corresponding Author: Suzan Demir Pektas, Department of Dermatology, Muğla Sıtkı Kocman University Faculty of Medicine, 48000, Mugla, Turkey.
T.: +90 2522115219 F.: +90 3123116768 E-Mail: suzandpektas@gmail.com

Background

PXE is a multisystem disease with an autosomal dominant inheritance pattern in 90% of the cases, or autosomal recessive [1]. Females are more commonly affected than males [2]. PXE is caused by mutations in the ABCC6 (ATP-binding cassette subfamily C member 6) gene [2]. Clinical manifestation occurs in skin, eyes, oral mucosa, gastrointestinal tract and the arteries [1]. We report two cases of PXE, emphasizing its main clinical aspects.

Case Report 1

Female patient, 48 years old reported the onset of asymptomatic yellowish-colored cobblestone-like papules and plaques in the cervical and axilla region. She had three intravitreal injections for treatment of choroidal neovascular membrane in ten years. She had a history of vision blurring. The patient did not have underlying diseases and denied familial cases of the same malady. At the dermatological examination, coalescent yellowish papules forming plaques distributed symmetrically in the cervical region and axilla were observed (Figure1A). The histopathological examination made evident calcified, distorted, and fragmented elastic fibers in the dermis with Verhoeff–van Gieson stain, compatible with the diagnosis of PXE (Figure 1 C, D). Fundus fluorescein angiography couldn't be performed due to patient's fluorescein allergy. BCVA was 0.2 (Snellen) in the right eye and counting fingers in the left eye. Slit-lamp examination of the anterior segment and intraocular pressures were within normal limits. Fundoscopic examination and optical coherence tomography examination revealed angioid streaks around the optic discs in both eyes and chorioretinal scar due to the choroid neovascularization in the left eye (Figure 2A, B). On other laboratory and cardiologic examination, were unremarkable. She has been made Dermatologic and Ophthalmologic monitoring.

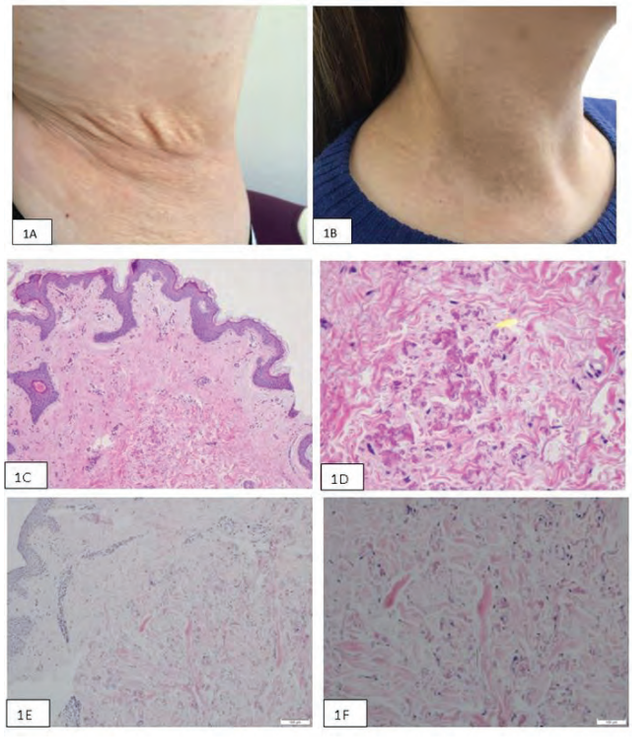

Figure 1. Coalescent yellowish papules forming plaques distributed symmetrically in the axilla of the first case (A); Coalescent yellowish papules forming plaques distributed symmetrically in the cervical region of the second case (B); At histological examination of the first case: Fragmented elastic fibers in the reticular dermis (x100) (C); Short, distorted basophilic elastic fibers (x400) (D); At histological examination of the second case: degenerative elastic fibers in subcutaneous tissue (X10) (E); Degenerative elastic fibers with elastic van Gieson stain (X200) (F).

Case Report 2

26 years old female patient presented to our clinic with asymptomatic yellowish-colored papules in her cervical region. At the dermatological examination, several brown colored macules and erythematous papules and coalescent yellowish papules in the cervical region were observed (Figure 1B). The patient did not have underlying diseases and denied familial cases of the same malady. The histological examination of papules revealed calcification and fragmentation of elastic fibers in the middle and deep dermal layers, confirming the diagnosis of PXE (Figure 1E, F).

BCVA was 1.0 in both eyes (Snellen). Slit-lamp examination of the anterior segment and intraocular pressures were within normal limits. Fundoscopic examination revealed AS around the optic discs and peau d'orange appearance in the temporal macula of both eyes (Figure 2C). Fluorescein angiography showed variable staining of the AS without leakage in any phase (window defect) and Comet signs in the mid-periphery of the retina (Figure 2D). Fundus autofluorescence showed distinct areas of hypo-autofluorescence corresponding to the AS with focal spots of increased autofluorescence alongside the AS (Figure 2E). SD-OCT showed breaks in Bruch's membrane with preservation of the overlying retina pigment epithelium (Figure 2F). Laboratory and cardiologic examination were unremarkable. Dermatologic and Ophthalmologic monitoring has been made.

Discussion

PXE is a genetic disorder of the connective tissue characterized by skin, ocular and vascular lesions [1, 2]. Asymptomatic skin manifestations, which are often the first clinical signs of PXE,

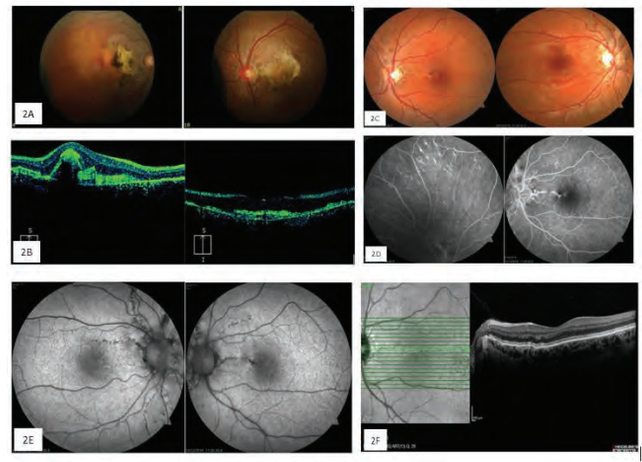

Figure 2. In ophthalmic examination of the first case: Color fundus photography of a patient with angioid streaks around the optic disc (A); chorioretinal scar due to the choroidoretinal neovascularization (B); In ophthalmic examination of the second case: Color fundus photo showing typical angioid streaks and peau d'orange appearance in the temporal of the macula (C); Window defect in fluorescein angiography due to atrophy of RPE adjacent to angioid streaks (D); Fundus autofluorescence (FAF) imaging shows areas of hypo-autofluorescence in the peripapillary area that are greater in size than the corresponding AS seen on color photography (E); SD-OCT findings of the patient showing breaks in Bruch's membrane with preservation of the overlying retina pigment epithelium. The yellow line indicates the location of the SD-OCT scan (F).

usually occur between the first and second decades of the patient's life [3]. The skin lesions typically consist of small, asymptomatic, yellowish, or skin-colored papules, which progressively coalesce into larger plaques and plaques due to cutaneous laxity [2, 3]. Skin alterations commonly appear during childhood and progress slowly and unpredictably during adulthood. They are initially located on the lateral and posterior regions of the neck. Flexural skin areas are frequently involved in the progression of the disease. Mucosal lesions of the oral cavity and genital area can also be detected and resemble cutaneous changes. Although the cutaneous lesions principally represent a cosmetic problem, they predict the risk for development of ocular and cardiovascular manifestations, with a considerable morbidity and occasional mortality [2, 3]. Because the lesions are thin and asymptomatic, the diagnosis is delayed [2]. In our two cases, the cutaneous lesions of PXE were asymptomatic. While the cutaneous lesions of PXE in the first patient were detected fourth decades of the patient's life, the cutaneous lesions of PXE in the second patient were detected second decades of the patient's life.

Ophthalmological features of PXE include primarily peau d'orange, comet lesions, angioid streaks, choroidal neovascularization (CNV), hemorrhages. Other ocular signs of PXE include chorioretinal atrophies, optic disk drusen and disciform scar [2, 3]. Peau d'orange is the earliest funduscopically visible alteration in patients with PXE, preceding the development of angioid streaks. Angioid streaks are the most obvious and consistent features of PXE fundus abnormalities. Approximately 85% of patients with PXE exhibit angioid streaks caused by dehiscence of the Bruch membrane, which appears thickened, calcified, and abnormally fragile. Angioid streaks are characteristic, but not pathognomonic. In later stages of the disease, an ingrowth of fibrovascular tissue through the defect may occur, giving way to secondary CNV and subsequent development of a disciform scar with subretinal fibrosis and atrophy [1, 2]. Secondary degenerative and hemorrhagic changes in the macula can frequently be found, leading to the severe reduction of visual acuity [1]. The vision blurring symptoms in our first case were due to the secondary CNV which was occurred as a complication of the dehiscence of the Bruch membrane. There were no visual complaints in our second case, where only angioid streaks, peau d'orange appearance, comet signs and breaks in Bruch's membrane which was visualized in OCT scans were detected. These patients, like in our case 2, should be examined much closely than other patients. Because Bruch membrane breaks may cause CNV and these membranes may cause severe vision blurring when untreated.

In the cardiovascular system, the calcification of artery walls of small and medium caliber is observed [1, 2]. The patients of PXE can also develop premature atherosclerosis with early acute myocardial infarcts [2, 3]. Alterations in lipoprotein composition and bleeding diathesis were found in plasma samples of PXE patients [2].

The diagnosis is clinical, associated with histopathological examination, which is characteristic and reveals fragmented and distorted elastic fibers in the reticular and deep dermis. These changes are more evident in the Verhoeff, Van Giesson and Calleja stains, specific for the elastic tissue [2, 3]. The clinic manifestation of skin lesions, ocular lesions, and histopathological findings in skin lesions in both of our cases were compatible with PXE.

Numerous systemic and dermatologic disorders could manifest clinical and histological features resembling classic PXE. Moreover, the absence of skin alterations does not exclude a diagnosis of PXE [2]. Cutaneous lesions of PXE-like phenotype have been described in association with vitamin-K dependent coagulation factor deficiency [4]. The histological finding is indistinguishable from classic PXE on light microscopy [4]. Other dermatologic disorders resembling PXE are cutis laxa, fibroelastolytic papulosis, PXE-like papillary dermal elastolysis, late-onset focal dermal elastosis, and perforating calcific elastosis [2]. The elastosis perforans serpiginosa, upper and mid-dermal elastolysis, papular elastorrhexis and linear focal elastosis, can manifest similar histological phenotypes as observed in PXE [2]. Clinical features are closely resembling PXE are also reported in association with inherited hemoglobinopathies [2].

Angioid streaks are also associated with a many of disorders, including Paget disease, hemoglobinopathies and Marfan [2]. These diseases should be considered in the differential diagnosis of the PXE patients with angioid streaks.

There is no specific or effective treatment in PXE. The therapeutical management is based on prevention and monitoring of complications associated with the disease [1]. Surgery for aesthetic improvement of cutaneous lesions is not routinely performed. However, significant progress has been made in the therapy of ocular complications [2, 5]. The diet supplemented with magnesium and vitamin K may extend the progression of the disease [1].

In conclusion, one must be aware of the need for early diagnosis, recognizing the typical cutaneous manifestations of the disease and better management of the associated complications when these are present [2].

Competing interests

No conflict of interest was declared by the authors.

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Case Report
JCAM

A Leukocytoclastic Vasculitis Case Due to Tenofovir Use

Tenofovir Kullanımına Bağlı Gelişen Lökositoklastik Vaskülit Olgusu

Tenofovir Induced Leukocytoclastic Vasculitis

A Haykir Solay[1], F Civelek Eser[1], EE Tutuncu[1], B Gencler[2], E.Ozturk Onder[3]
[1]Department of Infectious Disease and Clinical Microbiology, [2]Department of Dermatology,
[3]Department of Pathology, Diskapi Yildirim Beyazit Training and Research Hospital, Ankara, Turkey

Öz

Asiklik bir nükleotid analoğu olan tenofovir disoproksil fumarat, hepatit B tedavisi için 2008 yılında onaylanan antiviral bir ilaçtır. Bilinen en sık yan etkileri; bulantı, kusma, diyare, nefrotoksisite ve hepatotoksisitedir. Fakat cilt tutulumu olan yan etkiler nadir görülür. Kutanöz lökositoklastik vaskülit ise birçok ilaca bağlı olarak görülebilmekle birlikte tenofovirin indüklediği olgu daha önce hiç bildirilmemiştir. Burada, kronik hepatit B tanısı ile başlanan tenofovir tedavisinin dördüncü haftasında kutanöz lökositoklastik vaskülit gelişen bir olgu sunulmuştur.

Anahtar Kelimeler

Tenofovir; Lökositoklastik Vaskülit; Yan Etki

Abstract

The antiviral agent tenofovir disoproxil fumarate is an acyclic nucleotide analogue, which has been approved in 2008 for the treatment of hepatitis B. The most common side effects are nausea, vomiting, diarrhea, nephrotoxicity, and hepatotoxicity. Nevertheless, side effects with dermal involvement are rare. Cutaneous leukocytoclastic vasculitis can be induced by many drugs, but the tenofovir-induced case has not been reported previously. To best of our knowledge, we present the first case of tenofovir induced cutaneous leukocytoclastic vasculitis at the 4th week of treatment of chronic hepatitis B.

Keywords

Tenofovir; Leukocytoclastic Vasculitis; Side Effect

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Corresponding Author: Asli Haykir Solay, Department of Infectious Disease and Clinical Microbiology, Diskapi Yildirim Beyazit Training and Research Hospital, University of Health Sciences, Altindag, Ankara, Turkey. F.: +90 3125962793 E-Mail: aahaykir@hotmail.com

Introduction

Cutaneous leukocytoclastic vasculitis (LCV) is the inflammation of small blood vessels and may be secondary to drugs, infections, malignity, and connective tissue disorders [1].

Tenofovir is generally well tolerated. Most common side effects are related to the gastrointestinal system [2]. Nephrotoxicity and hepatotoxicity are less common. However, side effects, involving the skin, are rare [3]. In our knowledge, LCV case due to tenofovir has not been reported yet. Here, we present a case with LCV, which developed at the first month of tenofovir treatment.

Case Report

A 59 years-old male patient with chronic hepatitis B and cirrhosis admitted to our hospital with widespread itching, particularly at lower extremities. Medical history revealed that complaints of the patient started in the first month of tenofovir treatment, and no other medications were used. He had no complaint other than weakness, and itching; he had no fever, oral or genital ulcers, photosensitivity, arthralgia; and he did not take any herbal treatments. Physical examination revealed that heart beat was 82 bpm, blood pressure was 110/70 mmHg, respiratory rate was 16/min, and body temperature was 36.7 °C. There were no palpable lymph nodes. Hyperpigmented macular lesions of 0.5 cm in diameter were present in both lower extremities, particularly on anterior of tibias (Figure 1). Remaining physical examination findings were normal. Complete blood count, hepatic, renal and thyroid function tests, erythrocyte sedimentation rate and C-reactive protein were normal. Other laboratory findings are summarized in the table. Skin biopsy was performed after learning that previous antihistaminic treatment did not work, which was recommended by a dermatologist. Biopsy result was reported as mild, and partly moderate inflammatory infiltrate at perivascular localization in the subepidermal region beneath the epidermis, which included lymphocytes as well as high numbers of polymorphic nucleated leucocytes. Partial leukocytoclastic foci were observed, and capillary endothelium made explicit. Additionally, few numbers of extravasated erythrocytes were seen in the interstitium. These findings were compatible with LCV (Figure 2). Possible etiologies for LCV include infectious diseases, malignity, and collagen tissue disorders. Since he was on follow-up for chronic hepatitis B, HBV DNA was analyzed, but found negative. Other viral indicators were also negative (Table). Pathogen microorganisms were not found in stool, throat, and sputum cultures. Urine cultures were also negative. Thoraco-abdominal tomography did not reveal positive findings for infectious diseases or malignity. There were no pathological findings other than esophageal varices in upper gastrointestinal system endoscopy. Colonoscopy did not reveal findings of chronic inflammatory bowel disease or malignity. There was no finding regarding collagen tissue disorders. Serologic indicators for collagen tissue diseases and other vasculitis are presented in Table. Only cytoplasmic antinuclear antibody was found positive. The diseases that might be responsible for the etiology were all eliminated, and the only drug that patient used, tenofovir, was stopped and replaced with entecavir. Itching and cutaneous lesions regressed after two weeks of drug exchange, and no new lesions were observed.

Table. Laboratory findings, and viral and serologic markers after development of leukocytoclastic vasculitis due to tenofovir

	Value
Complete Urine Test	
Leukocyte, erythrocyte, protein	Negative
Viral Markers	
HBsAg (ELISA)	Positive
HBV DNA (PCR)	Negative
Anti-HCV (ELISA)	Negative
Anti-HIV (ELISA)	Negative
Anti-HAV IgM (ELISA)	Negative
Delta antibody (ELISA)	Negative
CMV IgM/IgG (ELISA)	Negative
Serologic Markers	
Rheumatoid factor (IU/L)	Negative
Anti-nuclear antibody	Negative
P-ANCA	Negative
C-ANCA	Positive

Discussion

Leukocytoclastic vasculitis is a small-vessel vasculitis that is characterized by neutrophilic infiltration in cutaneous superficial postcapillary venules. It may be idiopathic, and may also be related to infections, drugs, collagen tissue diseases, and malignities [1]. The case presented here had no suspected disease in the etiology.

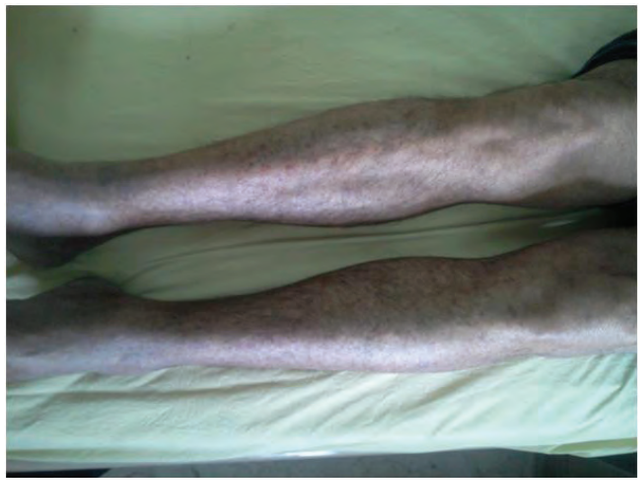

Figure 1. Multiple palpable purpuras in both lower extremities

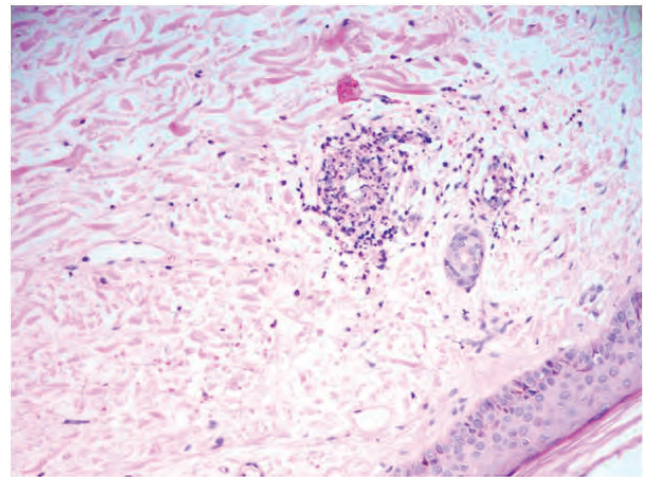

Figure 1. Lymphocytes and polymorphonuclear leukocytes in perivascular area releated to inflamation (HE X 200)

Small-vessel vasculitis are evaluated under two headings either antineutrophil cytoplasmic antibody (ANCA) positive or ANCA negative. These antibodies are autoantibodies that target lysosomal enzymes of neutrophils and grouped as c-ANCA that is against proteinase, and p-ANCA that is against myeloperoxidase [4]. ANCA positive small-vessel vasculitis are classified under four groups as microscopic polyangiitis, Wegener's granulomatosis, Churg-Strauss syndrome, and drug-induced vasculitis [5]. Drug-induced small-vessel vasculitis constitutes about 10%, and most commonly responsible drugs are penicillin, aminopenicillin, sulphonamides, allopurinol, quinolone, thiazides, hydantoin, and propylthiouracil [5]. They develop approximately after 7 to 21 days of drug initiation [6]. In our case, leukocytoclastic vasculitis has developed due to tenofovir, which has not been reported before and has developed one month after the initiation of the drug.

More commonly palpable purpura, especially in lower extremities, and less commonly urticarial, vesicular, nodular, and target-like lesions, livedoid pattern, and ulcerations are seen clinically [1]. Constitutional symptoms, arthralgia, myalgia, and other symptoms related to internal organ involvement are also seen in patients [7]. Itching is present rarely [8]. In this case, the main complaint of the patient was itching, and purpura. Tenofovir related leukocytoclastic vasculitis case has not been reported previously. Diagnose of this case was based on elimination of other causes, tenofovir being the only medication, and regression of lesions after changing this drug.

Tenofovir is a nucleotide analogue and widely used for chronic hepatitis B and antiretroviral treatment. It is a quite safe drug, but adverse events like nausea, vomiting, diarrhea, hepatotoxicity, nephrotoxicity, pancreatitis, Fanconi anemia, and diabetes insipidus were reported. Maculopapular rashes, urticarial rashes, vesiculopustular lesions, and lichenoid drug eruptions are rarely reported [3]. Also, tenofovir related itching is very rare [8]. The ideal way of treatment of cutaneous side effects related with tenofovir is not known. In a previous report of a case with skin eruptions, lesions were reported to regress ten days after the drug discontinuation [8]. In our case, no additional treatment was given, and itching complaint and lesions were resolved within two weeks after drug withdrawal. Increasing number or reports will help to constitute a common approach regarding the management of these cases.

Leukocytoclastic vasculitis is diagnosed by histopathological evaluation of the biopsy from the lesion, and additional laboratory tests should be performed for systemic involvement and its etiology.

Drugs might play a role in the etiology of LCV. Although it has not been reported before, LCV may develop on tenofovir use as can be seen in this case. One should be careful with the cutaneous lesions that develop during medication use and appropriate diagnostic methods, and the differential diagnoses should be kept in mind to manage the treatment.

Scientific Responsibility Statement

The authors declare that they are responsible for the article's scientific content including study design, data collection, analysis and interpretation, writing, some of the main line, or all of the preparation and scientific review of the contents and approval of the final version of the article.

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Ethical Responsibilities

No animal or human studies were carried out by the authors for this article.

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Clozapine Induced Enuresis Treated with Amitriptyline: A Case Report

Amitriptilin İle Tedavi Edilen Klozapine Bağlı Enüresis: Bir Olgu Sunumu

Clozapine Induced Enuresis Treated with Amitriptyline

Mehmet Emin Demirkol, Lut Tamam
Department of Psychiatry, Cukurova University Faculty of Medicine, Adana, Turkey

Öz

Tardiv diskinezi, genelde antipsikotik kullanımı sonrasında dopamin 2 reseptörlerinin uzun süreli blokajına bağlı olarak ortaya çıkan istemsiz hareketlerle karakterizedir. Aripiprazole bağlı nadiren gelişen tardiv diskinezi olgularında tedavi seçeneklerinden biri klozapindir. Klozapin eşsiz reseptör profili ile etki gücü ve yan etki anlamında diğer antipsikotiklerden ayrılır. Klozapin farklı mekanizmalar üzerinden enüresise neden olabilir. Klozapine bağlı enüresis tedavisinde farmakolojik seçenekler arasında desmopressin, oksibütinin, triheksifenidil, amitriptilin, aripiprazol, efedrin, verapamil bulunmaktadır. Hastanın psikiyatrik tanısı, seçenekler arasında bulunan ilaçların olası yan etkileri ilaç tercihi konusunda belirleyici olmaktadır.

Anahtar Kelimeler

Amitriptilin; Aripiprazol; Klozapin; Enüresis; Tardiv Diskinezi

Abstract

Tardive dyskinesia is characterized by involuntary movements which appear in connection with long-term blockage of dopamine 2 receptors following the use of antipsychotics. One of the treatment options in cases of tardive dyskinesia which occasionally induced by aripiprazole is clozapine. Clozapine is distinguished from other antipsychotics by its effective strength because of its unique receptor profile and its side effects. Clozapine may cause enuresis by various mechanisms. Among the pharmacological choices for the treatment of clozapine-related enuresis are desmopressin, oxybutynin, trihexyphenidyl, amitriptyline, aripiprazole, ephedrine, and verapamil. The patient's psychiatric diagnosis determines the choice of medication with their possible side effects.

Keywords

Amitriptyline; Aripiprazole; Clozapine; Enuresis; Tardive Dyskinesia

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Corresponding Author: Mehmet Emin Demirkol, Department of Psychiatry, Cukurova University Faculty of Medicine, Balcalı Hospital, 01160, Adana, Turkey.
GSM: +905355849684 E-Maeil: emindemirkol@gmail.com

Introduction

Tardive dyskinesia (TD) is characterized by involuntary athetoid, choreiform, ballistic dyskinetic movements which appear after Dopamine-2 (D_2) receptor blockage related to the long-term use of the first generation (conventional, typical) antipsychotics such as haloperidol or fluphenazine [1,2]. Among the risk factors for TD are advanced age, movement disorders present before the use of medication or a diagnosis of neurodegenerative disease, the use of lithium and the use of antipsychotics for more than six months [1,3]. After the discovery of the second generation (atypical) serotonin 2 ($5\text{-}HT_2$) receptor blocking antipsychotics such as quetiapine, risperidone, and olanzapine, the rate of occurrence of TD was partly reduced. Because of its partial agonistic activity, aripiprazole is distinct from other second generation antipsychotics and is known as an atypical antipsychotic [1]. The incidence of tardive dyskinesia in the population of non-geriatric users of second generation antipsychotics in a study was found to be 0.8% [1]. As well as this expected low level with the use of aripiprazole, there are also cases in the literature of dyskinetic movements among cases with persistent depression, bipolar disorder, obsessive compulsive disorder, schizoaffective disorder, and schizophrenia. The average daily dose of aripiprazole used with these patients was 10-20 mg [1,4].

Clozapine is an atypical antipsychotic with proven effectiveness in TD developing after the use of antipsychotics [5]. Although it has fewer extrapyramidal side effects, it causes more agranulocytosis, an increase in the risk of stroke, sedation, an increase in obsessive-compulsive symptoms, weight gain and sialorrhea compared with other antipsychotics. In addition to these side effects, 6-44.3% of patients who use clozapine complain of urinary system symptoms, especially enuresis [6]. Various hypotheses have been advanced to explain clozapine-related enuresis. These are urinary retention and overflow incontinence related to the antimuscarinic effect of clozapine, the cholinomimetic effect of clozapine, reduction in internal sphincter tone in connection with α-1 receptor blockage, the sedative effect of clozapine, and an increase in urinary retention relating to a lowering of the seizure threshold and constipation [6]. Among the options for the treatment of clozapine-related enuresis are reducing the dose of clozapine, behavioral recommendations and use of several different drugs (desmopressin, oxybutynin, trihexyphenidyl, amitriptyline, aripiprazole, ephedrine, and verapamil). Amitriptyline and aripiprazole are foremost among these treatments in patients with an affective component [6]. In this case report, we would like to report a clozapine induced enuresis which has been resolved by the addition of amitriptyline.

Case Report

A female patient who was 38 years old, a university graduate, a teacher and single, attended our outpatients' department accompanied by her family in October 2011, with complaints of irritability and sleeplessness. In her mental examination performed at that time, she was observed to have reduced care for herself, to have persistent ideas of persecution towards her husband and principal, to have emotions of anger and irritability. She was admitted to our hospital with a preliminary diagnosis of a schizoaffective disorder bipolar type manic episode. The patient stayed in our clinic for 36 days and a treatment of lithium 1200 mg/day and paliperidone 6 mg/day was started. She was discharged with full remission. In February 2012, she stopped taking the medication without the consent of her physician. One month after stopping the medication, she began to accuse herself and her family, to talk to herself, and to spend money unnecessarily. At that time she also had complaints of inappropriate laughing, an increase in sexual desire, and sleeplessness. As her complaints recurred, she was hospitalized again with the same diagnosis. The patient had auditory hallucinations and remained in our clinic for 32 days. She was prescribed lithium 1200 mg/day and aripiprazole 30 mg/day. She responded well to this treatment and discharged one month later with full remission. Between 2012 and 2016, the patient received her medication as suggested and has regularly been followed up in the outpatient clinic. She was able to continue her job as a teacher, and she experienced no manic, depressive or psychotic symptoms. At the beginning of June 2016, she began to have sleeplessness, twitches in her eyes, contraction, numbness in her face, contraction in her mouth and involuntary movement of her lips. Thinking that the complaints would recede by themselves over time, she did not seek medical help. Later she went to ophthalmology and neurology. The patient was referred to our outpatients' department by neurology with suspected TD, and was admitted to our clinic.

In her initial psychiatric evaluation at the time she was admitted to our clinic, she did not have thoughts of irritability, suspiciousness, depressive elements, or grandiose thoughts. She had no visual or auditory hallucination. Her memory and orientation were normal. Involuntary movements were observed around her mouth and eyes. She scored 26/42 on the Abnormal Involuntary Movement Scale (AIMS). After neurological consultation, it was decided that these involuntary movements could be consistent with TD, and it was recommended that drugs which could give rise to this should be discontinued or changed. No further recommendations were made after ophthalmology consultation. Being female, having mood components, taking lithium and taking antipsychotics for more than six months were considered as risk factors for TD. In the first laboratory examination, her hemogram, liver, renal, thyroid functions and fasting blood glucose were within normal limits. Her blood lithium level was 0.71 mEq/ liter. No pathology was found in her brain magnetic resonance imaging (MRI). Assessment of her medication history revealed that her current dyskinetic movements could be related to aripiprazole, and that her use of lithium might contribute to this. As the patient had been benefitting from lithium for approximately six years, it was first planned to discontinue aripiprazole and begin clozapine, vitamin E, and diazepam. Before initiation of clozapine, hemogram, electroencephalography, and echocardiography tests were conducted and found to be normal. The clozapine dose has been increased gradually. The patient's involuntary eye movements decreased (AIMS: 19/42) in the third week of treatment when the clozapine dose was raised to 250 mg/day. At this time her blood lithium level was 0.82 mEq/ liter. In the fourth week of clozapine treatment, the patient began to have complaints of enuresis nocturna. After a few days, she began to have daytime urinary incontinence. Hemogram and urine tests were within normal limits. Neurologi-

cal, internal medicine and urological consultations' results eliminated other possible causes of enuresis (i.e., urinary infection, diabetes mellitus, renal failure, epilepsy). Lithium and diazepam doses were reduced and terminated. Despite the cessation of all other drugs, enuresis continued. This has shown us possible association between clozapine and patient's enuresis. One week after this, the complaint of enuresis was continuing, and it was decided to start amitriptyline 25 mg/day. Urinary incontinence ceased after the second day of amitriptyline treatment, and after a follow-up of seven days, the amitriptyline dose was stopped. Four days after this, the complaint of enuresis began again, and the patient was once more prescribed amitriptyline 25 mg/day. In the second day of treatment, the enuresis complaint ceased again. The patient was discharged from hospital after a stay of 99 days. In her follow up she was in good condition about tardive dyskinesia (AIMS:16/42) and enuresis after six months with a treatment regimen of clozapine 250 mg/day and amitriptyline 25 mg/day.

Discussion

Clozapine is distinguished from other antipsychotics by its high D_1 and low D_2 receptor activity. It has been suggested that D_1 receptor affinity is effective in the treatment of TD. Clozapine has side effects like agranulocytosis, hypotension, seizures, constipation, weight gain, sialorrhea and enuresis [5]. Recent publications have shown that 6-44.3% of patients using clozapine have urinary system symptoms, especially enuresis [6]. Presence of enuresis contributes to poor medication adherence so has deleterious effects on the prognosis of psychosis [7]. Although there are various treatment options (desmopressin, oxybutynin, trihexyphenidyl, amitriptyline, aripiprazole, ephedrine, and verapamil) for the complaint of enuresis induced by clozapine, amitriptyline 25 mg/day was preferred in our case because of the existing diagnosis of schizoaffective disorder and the recurring depressive symptoms.

Amitriptyline, a tricyclic antidepressant, has antimuscarinic, antihistaminic and antiadrenergic effects. Amitriptyline is effective in the treatment of enuresis and clozapine-induced sialorrhea because of its anticholinergic effect. It also increases vasopressin secretion and shortens the period of REM sleep, these effects contribute to the treatment of enuresis. Adding amitriptyline to clozapine treatment can give rise to anticholinergic side effects, lowering the seizure threshold and sedation. So adding low doses of amitriptyline to clozapine treatment can be rational. Our case, in which the complaint of enuresis ceased from the second day of amitriptyline treatment, is the second in the literature in which amitriptyline was beneficial in the treatment of clozapine induced enuresis [7].

In conclusion, this case report presents a treatment option with amitriptyline for clozapine induced enuresis cases but needs to be supported by further studies.

Competing Interests

The authors declare that they have no competing interests

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A Rare Cause of Renal Atrophy: Subcapsular Collection Presented as a Huge Perirenal Complex Cyst

Renal Atrofinin Nadir Bir Sebebi: Dev Perirenal Kompleks Kist Şeklinde Prezente Olan Subkapsüler Koleksiyon

Subcapsular Collection Presented as Perirenal Cyst

Abidin Kılınçer[1], Emre Ünal[2], Orhan Yıldız[3]
[1]Department of Radiology, Zile State Hospital, Zile, Tokat,
[2]Department of Radiology, Hacettepe University School of Medicine, Sıhhiye, Ankara,
[3]Department of Urology, Zile State Hospital, Zile, Tokat, Turkey

Öz

Renal subkapsüler koleksiyonlar tedavi edilmezse renal atrofi ile sonuçlanabilir. Tekrarlayan hemoraji epizodları koleksiyonun karmaşık görünümlü olmasına neden olduğu için kronik dönemde tanı koymak oldukça zor olabilir. Biz bu yazımızda 44 yaşındaki bir erkek olguda böbrek atrofisinin nadir bir sebebini sunuyoruz.

Anahtar Kelimeler

Atrofik Böbrek; Kompleks Kist; Subkapsüler Koleksiyon; MRG; BT

Abstract

If left untreated, renal subcapsular collections may result in renal atrophy. The diagnosis could be challenging at chronic stage due to recurrent episodes of hemorrhage which is the reason for the complex appearance of the collection. Herein we report a rare cause of renal atrophy in a 44-year-old man.

Keywords

Atrophic Kidney; Complex Cyst; Subcapsular Collection; MRI; CT

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Corresponding Author: Abidin Kılınçer, Department of Radiology, Zile State Hospital, Zile, 60400, Tokat, Turkey.
T.: +90 3563175098 GSM: +905536591317 E-Mail: akilincer@yahoo.com

Introduction

The contents of the renal subcapsular collections may vary depending on the etiology [1,2]. Subcapsular collections may induce significant pressure on renal parenchyma due to boundary effect of renal capsule. In the setting of renal vessel compression, one may identify the new onset of increased blood pressure due to activation of the renin-angiotensin system, which is also called as Page kidney [2,3]. However, in the case of long standing compression, the kidney may undergo atrophy and imaging based evaluation could be challenging. Herein we report a rare case of chronic renal subcapsular collection presented as a huge complex cystic mass resulting in significant right kidney atrophy.

Case Report

A previously healthy 44-year-old man was admitted to our hospital with a two days history of right back pain radiating through to right inguinal region. He was afebrile, and his blood pressure was within the normal range. There was no history of trauma. The urinary test revealed lack of hematuria. Serum creatinine level was normal (0.76 mg/dL). Abdominal radiography demonstrated a calcified ovoid mass in right flank (Figure 1). Ultrasonography revealed the cystic nature of the lesion with fluid-fluid level (Figure 2). Abdominal CT examination demonstrated a peripherally calcified, huge (sized 10 x 13 cm) cystic mass in right renal fossa (Figure 3). The right kidney was absent however there was a non-specific soft tissue density with a few punctate calcifications between the cystic mass and inferior vena cava (Figure 3). The left kidney was unremarkable except for hypertrophy. The right adrenal gland and urinary bladder were normal. Contrast-enhanced MRI examination was suggestive of right renal atrophy, however, neither renal artery nor renal vein were apparent on postcontrast images. The extension of the cyst was indicative of subcapsular origin (Figure 4). The patient underwent surgery, and histopathological examination revealed collagenized and calcified cyst wall associated with atrophic renal parenchyma and pelvicalyceal system. There was no sign of granulomatous reaction in histopathological examination, and also acid-fast bacillus staining was negative for tuberculosis. Urine culture test and chest radiograph were also unremarkable.

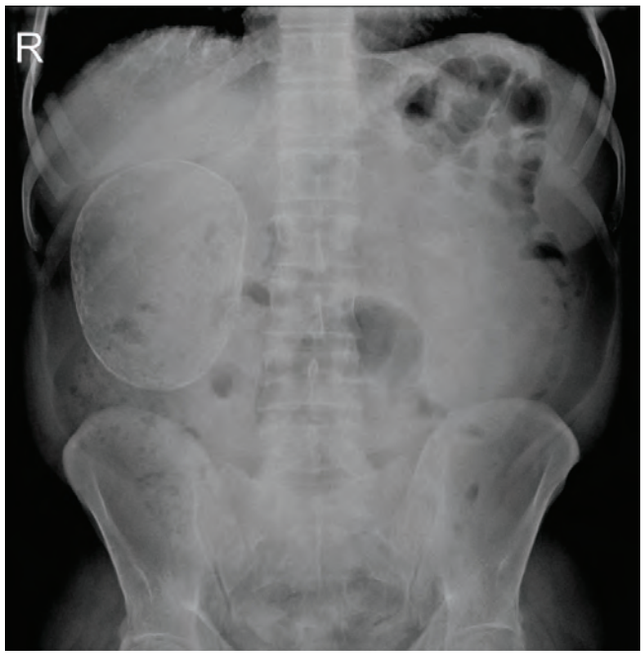

Figure 1. Abdominal radiography demonstrates a peripherally calcified ovoid mass in the right flank.

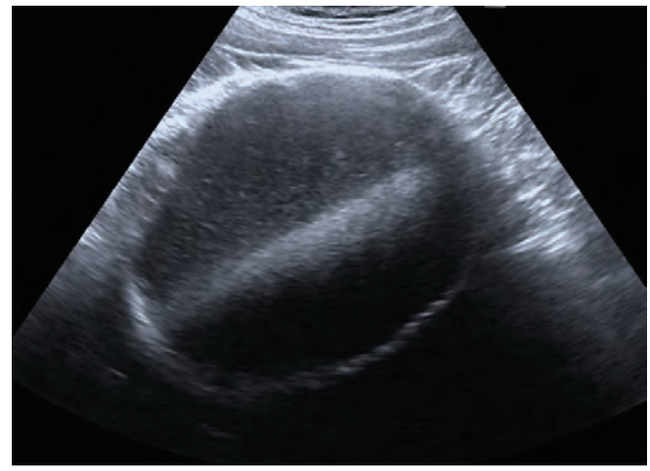

Figure 2. Gray scale sonogram reveals fluid-fluid level within the cyst lumen.

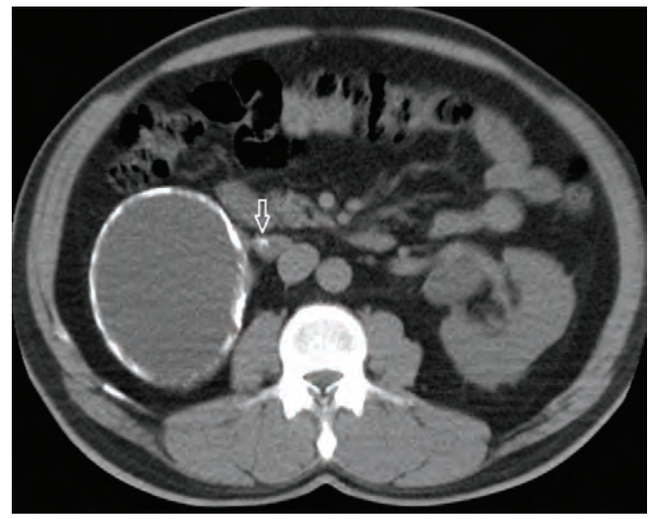

Figure 3. Axial unenhanced CT image shows nonspecific calcification containing soft tissue density located between the cystic mass and inferior vena cava (arrow).

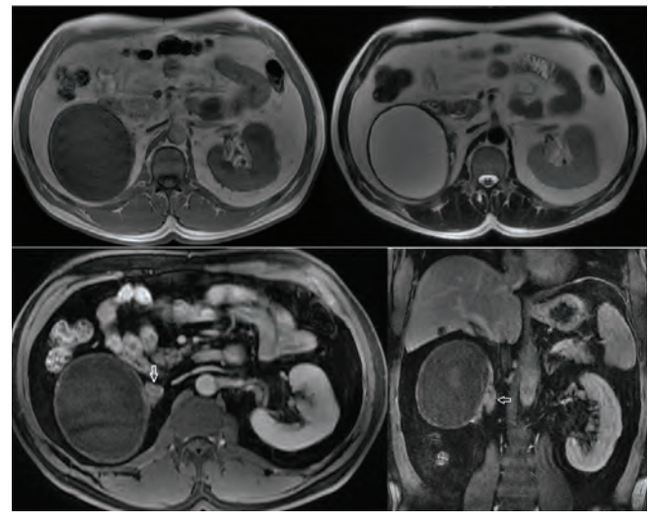

Figure 4. Axial T1 (a) and T2 (b) weighted MR images reveal lack of hemorrhage and debris within the cyst. Axial (c) and coronal (d) post-contrast fat saturated T1 weighted MR images demonstrate contrast enhancement adjacent to the cyst compatible with atrophic right kidney (arrows). The extension of the cyst indicates subcapsular origin (a-d).

Discussion

Herein reported case indicates that renal subcapsular collections may result in significant parenchymal atrophy at late stage. Compensatory hypertrophy of the contralateral kidney also suggests chronic state of the insult. The main limitation for further conclusion is that we were not able to evaluate previous radiological examinations of the patient. The patient's presenting symptom was newly onset of back pain. Although back pain is a nonspecific complaint that could be seen in various conditions, one may suggest complication of an existing cyst. However pathological examination revealed lack of hemorrhage and pus. Therefore, complication of a cyst was less likely the diagnosis. Also, the definition of "collagen-rich cyst wall" in pathologic report points out the renal capsule. Collagen is reported to be found in ultrastructure of the renal capsule as opposed to renal cysts which are collagen free. However, the underlying pathophysiology of our case remains unclear. Communication of subcapsular collection with the lymphatic system could be questioned as a triggering factor for fluid layering. However, pathological examination revealed lack of such association as well. The presenting symptom could be related to another cause (i.e., lumbar disc hernia or myalgia), and the renal atrophy could be incidentally diagnosed.

A hematoma is the most frequent type of renal subcapsular collection and may occur following trauma, anticoagulant therapy, and interventional procedures [1, 4-6]. Renal subcapsular pseudocyst may also be encountered as a complication of acute pancreatitis [3]. A urinoma can also be the underlying cause of subcapsular collection particularly in the setting of obstructive uropathy (i.e., ureteral stone, retroperitoneal fibrosis, bladder cancer) complicated by forniceal rupture [7,8]. On the other hand, trauma may also result in subcapsular urinoma as well [2]. In case of bilateral renal subcapsular collection presence of renal parenchymal diseases (i.e., membranoproliferative and focal-segmental glomerulonephritis) could be questioned [9,10]. In our case, neither imaging findings nor clinicopathological signs were able to reveal the underlying cause of renal subcapsular collection.

Lin et al. reported a case of tuberculosis autonephrectomy presented with a huge cystic mass with lack of normal appearing right kidney [11]. The cyst had calcified wall, and imaging findings were similar to our case. However, the urine culture test was positive for Mycobacterium tuberculosis in their case which is contrary to our findings. Moreover, the presence of compressed and atrophic kidney seen in our case is unusual for tuberculosis autonephrectomy.

In conclusion, renal subcapsular collections may result in renal atrophy, and the underlying cause may remain unknown particularly in patients presented at late stage. Presence of atrophic kidney adjacent to cystic mass indicates capsular-subcapsular origin of the cyst.

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Author contributions

Concept – A.K., E.Ü.; Design - A.K.; Supervision - A.K., E.Ü.; Resource - A.K., O.Y.; Materials - A.K., O.Y.; Data Collection and/or Processing - A.K., O.Y.; Analysis and /or Interpretation - A.K., E.Ü.; Literature Search - A.K.; Writing - A.K., E.Ü.; Critical Reviews - A.K., E.Ü.

Informed Consent

Written informed consent was obtained from the patient who participated in this study.

Conflict of Interest

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Malignant Nodular Hidradenoma of the Scalp: A Case Report

Kafa Derisinde Malign Nodüler Hidradenom

Malignant Nodular Hidradenoma of Scalp

Kadir Balaban[1], Murat Şedele[1], Alper Sayiner[2]
[1]Pathology Department, Health Science University, Antalya Education and Research Hospital, Antalya,
[2]Pathology Department, Şanlıurfa Mehmet Akif İnan Education and Research Hospital, Şanlıurfa, Turkey

Öz

Malign nodüler hidradenoma (MNH) nadir bir deri eki tümörüdür. Kafa derisi bu tümör için alışılmadık lokalizasyonlardan biridir. Bu çalışmada, kafa derisinde yavaş büyüyen kitle ile başvuran ve histopatolojik değerlendirme sonucu malign nodüler hidradenom tanısı alan 80 yaşında kadın hastayı sunmayı amaçladık. MNH mümkün olduğu kadar erken tanı alması ve cerrahi olarak geniş rezeksiyon yapılması gereken agresif bir tümördür. Buna karşın erken rekürrens sık olduğundan yakın klinik takip gerektirir.

Anahtar Kelimeler

Deri Eki Tümörü; Kafa Derisi; Karsinom; Malign Nodüler Hidradenom

Abstract

Malignant nodular hidradenoma (MNH) is a rare sweat gland tumor of the skin. The scalp is an uncommon site of occurrence. We, herein, present an 80-year-old woman whose slow-growing subcutaneous nodule on her scalp was diagnosed as malignant nodular hidradenoma after histopathologic examination. MNH is an aggressive tumor and should be diagnosed and excised as early as possible. Wide local excision is the treatment of choice. As early recurrence is common despite the wide excision close follow-up is necessary.

Keywords

Carcinoma; Malignant Nodular Hidradenoma; Scalp; Skin Appendage.

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Corresponding Author: Alper Sayiner,Mehmet Akif İnan Eğitim ve Araştırma Hastanesi Patoloji Kliniği, Haliliye, Şanlıurfa, Turkey.
GSM: +905357896369 E-Mail: alpersayiner@yahoo.com

Introduction

Malignant nodular hidradenoma (MNH) is an infrequent, highly malignant, primary skin tumor which may have both apocrine and eccrine variants [1]. Several synonyms have been described in the literature, like malignant clear cell myoepithelioma, malignant acrospiroma, clear cell hidradenocarcinoma, clear cell eccrine carcinoma, clear cell hidradenoma, solid-cystic hidradenoma and eccrine acrospiroma. Occurrence in the scalp is rare but well-known [2]. Most reported cases are in elderly individuals over 50 years of age, though they may occur at any age. MNH has a very poor prognosis, high recurrence and a high rate of metastases [3]. We presented one case of MNH managed at our clinic.

Case Report

An 80-year-old woman presented to our hospital with an enlarging painless nodular mass on the scalp. Gross examination showed 3 cm in size, pinkish, firm, nodular mass.

Histopathological examination showed tumor cells with clear cytoplasm arranged in nodules infiltrating the superficial dermis with comedonecrosis (Figure 1A). There were frequent mitotic figures (Figure 1B). The findings were suggestive of MNH. Many of the tumor cells were positive for high molecular weight cytokeratin (Figure 1C) and CAM5.2; and negative for BER-EP4, CD10, Androgen receptor, Estrogen receptor and p63. Ki67 was stained in 30% of neoplastic cells (Figure 1D).

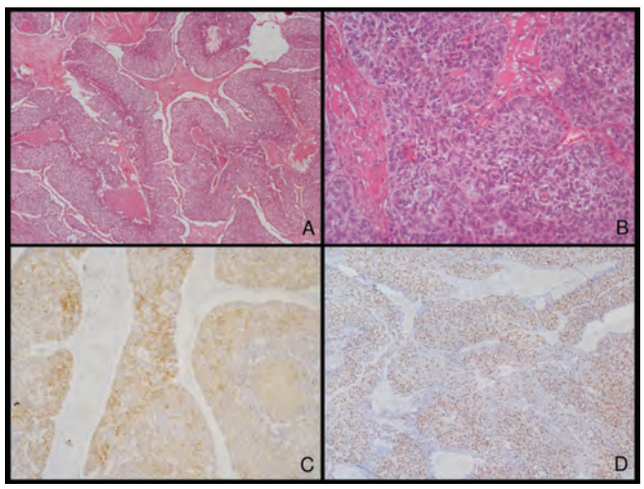

Figure 1. Tumor cells with clear cytoplasm arranged in nodules with comedonecrosis (H&E, x40)(A), Frequent mitotic figures in neoplastic cells (H&E, x200)(B), High molecular weight cytokeratin (HMWCK) positivity in neoplastic cells (HMWCK, x100)(C), Ki67 staining in 30% of neoplastic cells (Ki67, x100)(D).

Discussion

Tumors of the epidermal appendages are historically classified into four groups that exhibit histologic features analogous to hair follicles, sebaceous glands, apocrine sweat glands, and eccrine sweat glands [4]. Sweat gland tumors are mostly benign. Primary eccrine carcinomas are rare tumors and make up less than 0.01% of all skin cancers. Hidradenocarcinoma accounts for approximately 6% of malignant eccrine tumors and accounts for less than 0.001% of all tumors [5].

Malignant nodular hidradenomas are rare, and usually malignant from their inception; however, malignant lesions may arise from benign nodular hidradenomas. While nodular hidradenomas are typically well demarcated, malignant nodular hidradenomas are usually larger, asymmetrical, and show invasion into the surrounding tissue [4]. Nuclear changes may be absent, slight, or moderate [6]. The MNH displays atypical mitosis, necrosis, and angiolymphatic invasion. There are nodular or lobulated architecture of clear cells with glycogen containing cytoplasm and tubular or ductal structures. There may be a large or small representation of cells showing squamoid differentiation. However, a diversity of cell types, such as polygonal cells, clear cells, and spindle cells may be seen [1,2]. The cells express the high molecular weight cytokeratins CK5/6 and CK7, as well as p63, androgen receptor, estrogen receptor, and sometimes Her-2/neu. Ki-67 and p53 staining may be useful histologic parameters [3,6]. Even after the complete excision MNH has a potential for local recurrence, tends to metastasize, and often cause death [7]. In a report of seven cases from Italy, six patients died within 15-45 months of diagnosis [1]. Wide local excision is the treatment of choice with clearances of 2 cm recommended. Selective lymph node dissection is often used. The value of adjuvant radiotherapy has not been confirmed [3]. As early recurrence is common despite the wide excision close follow-up is mandatory [2].

Competing Interests

The authors declare that they have no competing interests

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Pneumothorax: An Important Cause of Anxiety

Pnömotoraks: Önemli Bir Anksiyete Nedeni

Pneumothorax

Banu Yoldaş, Figen Türk, Soner Gürsoy
Izmir Dr. Suat Seren Chest Diseases and Thoracic Surgery, Training and Education Hospital, Konak, Izmir, Turkey

To the editor:

We read the article of Yazkan et al. with great interest (1). Spontaneous pneumothorax is a common disease effecting escipecially young man who live their life more active. Various treatment methods have been offered in the literature. Some advice observation, simple aspiration as an initial therapy (2-4). But the risk of recurrence is a well known fact proven with the literature. All patients are informed in this direction. However in some patients this causes anxiety which results in a large number of unneccessary hospital applications. Yazkan et al. highlighted the importance of this topic in their series.

A 17 years old male patient was admitted to our hospital due to left sided spontaneous pneumothorax in 2015. After this date he applied to hospital 5 times without a serious complaint just because his anxiety. In 2017 he had a controlateral spontaneous pneumothorax treated with tube thoracostomy. After this time he applied two more times in a month. He asked for a computed tomography to different doctors and had three tomographies during this time. This patient is a good example of anxiety due to spontaneous pneumothorax. Out of such patients, we support the idea of Yazkan et al. marking “new surgical indication for spontaneous pneumothorax causing recurrent anxiety”. Escipecially thoracoscopic surgery might be a preferred approach in the first episode of spontaneous pneumothorax in young patients.

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Corresponding Author: Banu Yoldas, Izmir Dr. Suat Seren Chest Diseases and Thoracic Surgery, Training and Education Hospital, Konak, Izmir, Turkey.
T.: +90 2324333333 E-Mail: banuaktin@yahoo.com

Palivizumab Prophylaxis in Respiratory Syncytial Virus Epidemia; Neonatal Intensive Care Unit Experience

Respiratuar Sinsisyal Virus Salgınında Palivizumab Profilaksisi; Yenidoğan Yoğun Bakım Ünitesi Deneyimi

Palivizumab Prophylaxis

Fatma Hilal Yılmaz, Nazlı Dilay Gültekin, Hüseyin Altunhan
Yenidoğan Bölümü, Necmettin Erbakan Üniversitesi, Meram Tıp Fakültesi, Konya, Türkiye

To the editor:

Respiratory Syncytial Virus (RSV), which is an RNA virus with negative polarity, single helical and shielded form, belonging to the Paramyxovirus family, is one of the important reasons of Lower Respiratory Tract Infections in children below two years of age and in infants where they are prone to relapse [1]. The diseases progress in an extremely infectious manner, and may stay alive for nearly 1 hour in hands, 6 hours in secretions, 7 hours on hard surfaces; and the transmission mostly occurs via contact and droplets. According to the data received from Disease Control and Protection Centers, RSV is responsible for 2,3% of the newborn deaths [2]. RSV causes seasonal epidemic diseases in the whole world. The epidemics in the Mild Temperate Zone, where our country is also located, generally start as outbreaks in November-April and reach the highest level in January or February. Premature babies, children with Down Syndrome accompanied or not accompanied by neuromuscular diseases and cardiac diseases, patients with congenital or acquired immune defects are especially the candidates for serious infections as a high-level risk group. It has been reported in previously conducted studies that being premature alone is related to serious RSV infection at a significant level. Clinical findings vary according to the age and whether the infection is the first one or a relapsing one. Especially in newborns, they appear with the changes in chemoreceptor sensitivity, apnea and with the mechanism with which the reflex apnea is activated [3].

No vaccinations have been developed yet against this agent, which has high-level mortality and morbidity rates in the risk groups. Palivizumab, which is used in prophylaxis, is a humanized monoclonal antibody. It inhibits the RSV replication in Lower Respiratory Tract by preventing the fusion of RSV into respiratory epithelium cells and avoiding the syncytium formation [4]. The RSV infection diagnosed five (3-7) days after hospitalization is described as “Nosocomial RSV Infection”. When there are additional two or more cases in addition to the first case in a Newborn Intensive Care Unit, the existence of Nosocomial RSV Epidemic is considered.

A 23-day-old female patient was accepted in our unit with the complaints of respiratory problems and cough, and there was a pneumonic infiltration in the right paracardiac area in the front-rear chest radiography. It was determined by listening that the patient, who was tachypneic and dyspneic, had crepitant rales, especially in the right hemithorax. The hemogram, C-reactive protein and procalcitonin values of the patient were within normal ranges. The RSV A and B were found as positive in the viral swab panel of the patient. The patient was isolated and followed-up with supportive treatment, and was discharged with recovery from our unit on the 15th day of her hospitalization. 13 days after this patient was accepted in our unit, another patient who was born at 32 weeks and who was 21 days old showed apnea and respiration problems. The RSV fast antigen test was reported as positive. Three days after this case, a patient who was on the postnatal 122nd day and who was born at 26 weeks and diagnosed with bronchopulmonary dysplasia presented with cough complaints. The RSV fast antigen test was found to be positive. The patients were isolated, and the treatments continued with follow-ups. Upon this situation, which was considered as an RSV Epidemic, Palivizumab (15mg/kg/dose) was applied to 12 patients who were considered to be at risk. No new cases were detected after this application.

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Corresponding Author: Fatma Hilal Yılmaz, Yenidoğan BD, Necmettin Erbakan Üniversitesi, Meram Tıp Fakültesi, 42060, Konya, Türkiye. Tel: +90 3322236697 F.: +90 3322236182 E-Mail: f.h.yilmaz@hotmail.com